Primary Biliary Cholangitis; Acknowledged truths?

Now cholangitis and not cirrhosis

Progressive autoimmune disease

Female preponderance

Associated with cholestatic LFTs, elevated IgM, positive AMA.

Fatigue and itch are common symptoms

Approx. 15% cases present with advanced liver disease

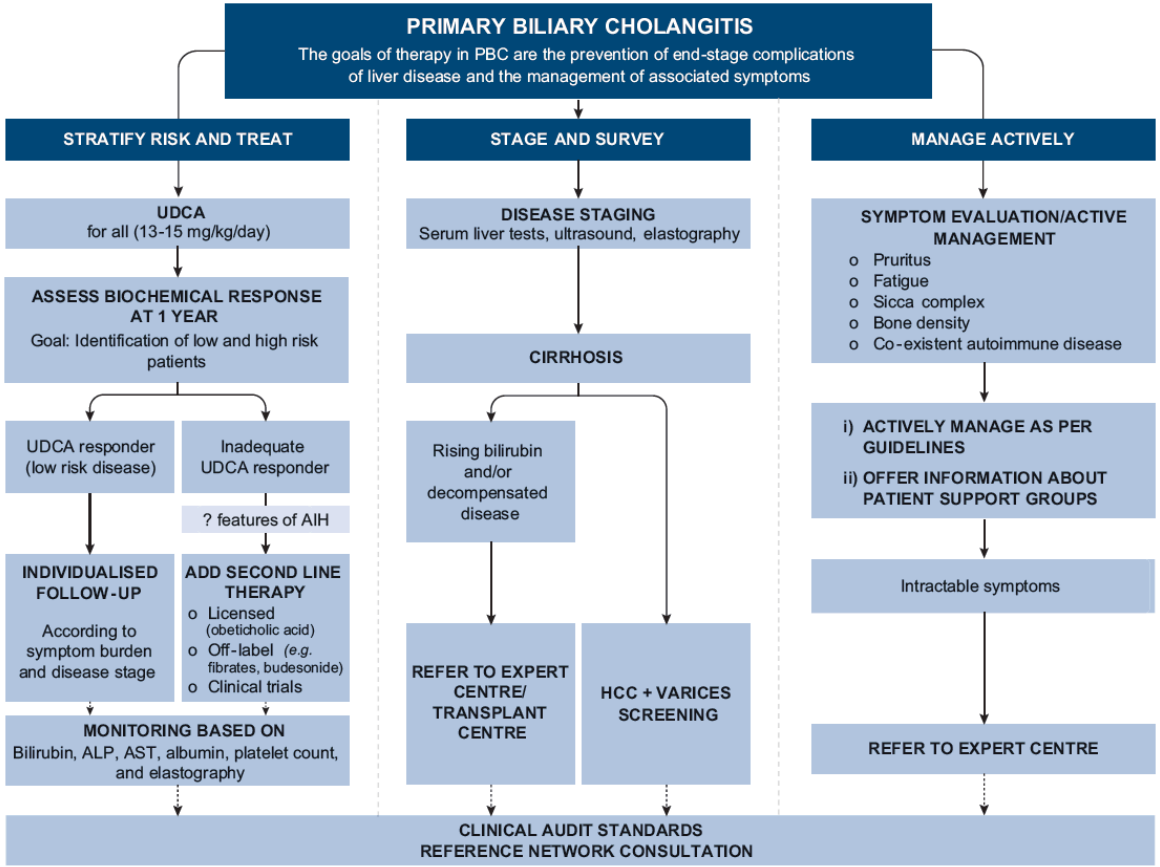
Ursodeoxycholic acid (10-15mg/kg) is standard of care

No curative therapy exists

JOURNAL OF HEPATOLOGY

EASL Clinical Practice Guidelines: The diagnosis and management of patients with primary biliary cholangitis☆

European Association for the Study of the Liver*



JOURNAL OF HEPATOLOGY

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Table 6. Rational approaches to risk stratification in PBC.

Level of applicability	Prognostic tools
High **(High applicability, robust validation)**	• On-treatment ALP and bilirubin-based assessment of response to UDCA using either qualitative or quantitative tools • Baseline (early *vs.* advanced) disease stage as defined by elastography, serum levels of bilirubin and albumin, or histology
Moderate **(High applicability, further validation pending)**	• LSM by elastography • APRI • ELF test
Indeterminate **(Limited applicability and/or validation)**	• Age, gender and symptom profile • PBC-specific ANA • Degree of interface hepatitis and ductopenia • Novel histological scoring systems • Direct measurement of portal pressure

ALP, alkaline phosphatase; UDCA, ursodeoxycholic acid; LSM, liver stiffness measurement; APRI, aspartate aminotransferase/platelet ratio index; ELF, enhanced liver fibrosis; PBC, primary biliary cholangitis; ANA, antinuclear antibodies.

Table 5. Assessing response to UDCA therapy in PBC.

Qualitative binary definitions	**Time (months)**	**Treatment failure**
Rochester [101]	6	ALP ⩾2× ULN or Mayo score ⩾4.5
Barcelona [62]	12	Decrease in ALP ⩽40% and ALP ⩾1× ULN
Paris-I [63]	12	ALP ⩾3× ULN or AST⩾ 2× ULN or bilirubin >1 mg/dl
Rotterdam [102]	12	Bilirubin ⩾1× ULN and/or albumin <1× ULN
Toronto [98]	24	ALP >1.67× ULN
Paris-II [104]	12	ALP ⩾1.5× ULN or AST ⩾1.5× ULN or bilirubin >1 mg/dl
Ehime [103]	6	Decrease in GGT ⩽70% and GGT ⩾1× ULN
Continuous scoring systems	**Time (months)**	**Scoring parameters**
UK-PBC [107]	12	Bilirubin, ALP and AST (or ALT) at 12 mo. Albumin and platelet count at baseline
GLOBE [106]	12	Bilirubin, ALP, albumin, and platelet count at 12 mo. Age at baseline

ALP, alkaline phosphatase; ULN, upper limit of normal; AST, aspartate aminotransferase; GGT, gamma-glutamyltranspeptidase.

Major Hepatic Complications in Ursodeoxycholic Acid-Treated Patients With Primary Biliary Cholangitis: Risk Factors and Time Trends in Incidence and Outcom

Maren H. Harms, MD[1], Willem J. Lammers, MD, PhD[1], Douglas Thorburn, MD, PhD[2,3], Christophe Corpechot, MD, PhD[4], Pietro Invernizzi, MD, PhD[5], Harry L.A. Janssen, MD, PhD[6], Pier M. Battezzati, MD, PhD[7], Frederik Nevens, MD, PhD[8], Keith D. Lindor, MD, PhD[9,10], Annarosa Floreani, MD, PhD[11], Cyriel Y. Ponsioen, MD, PhD[12], Marlyn J. Mayo, MD, PhD[13], George N. Dalekos, MD, PhD[14], Tony Bruns, MD, PhD[15], Albert Parés, MD, PhD[16], Andrew L. Mason, MD, PhD[17], Xavier Verhelst, MD, PhD[1] Kris V. Kowdley, MD, PhD[19], Jorn C. Goet, MD[1], Gideon M. Hirschfield, MD, PhD[20], Bettina E. Hansen, MSc, PhD[1,6] and Henk R. van Buuren, MD, PhD[1] on behalf of the Global PBC Study Group

The American Journal of GASTROENTEROLOGY

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Table 3. Predictive value of response criteria for occurrence of hepatic complications

Response criteria	HR	95% CI	*P* value
GLOBE-score	6.046	4.773–7.660	<0.001
Paris-I	5.024	3.856–6.546	<0.001
Paris-II	4.654	3.270–6.622	<0.001
Rotterdam	4.397	3.295–5.866	<0.001
Toronto	3.057	2.285–4.090	<0.001
Barcelona	1.711	1.274–2.296	<0.001

CI, confidence interval; HR, hazard ratio.

Biochemical response to UDCA was defined by previously reported criteria with a threshold defining response vs. non-response, calculated by univariate Cox regression analysis. Criteria were calculated after 1 year of follow-up, Toronto criteria were calculated after 2 years of follow-up.

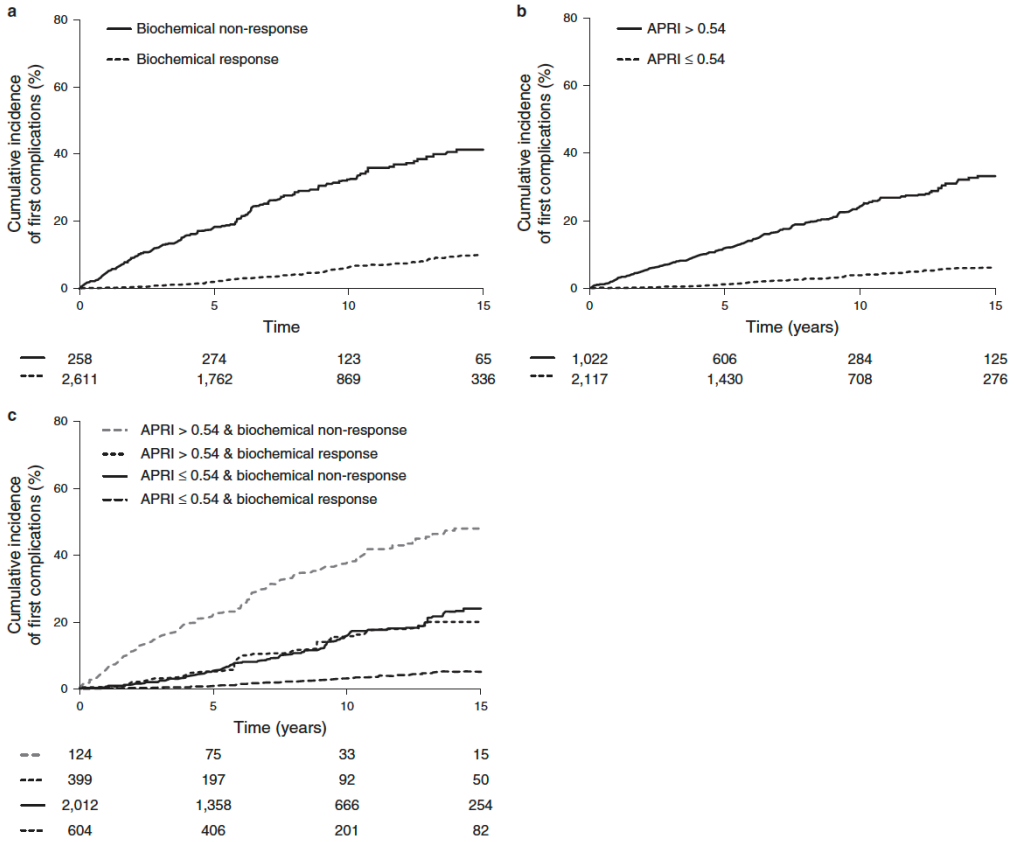

Figure 2. Cumulative incidence of hepatic complications according to biochemical response and aspartate aminotransferase to platelets ratio index (APRI) score. Kaplan–Meier estimates, stratified according to biochemical response (which was based on the age-dependent thresholds of the GLOBE score) and APRI score above or below the threshold of 0.54 are shown. The GLOBE score and the APRI score were calculated after 12 months of follow-up. *T*=0 represents the time after 12 months of follow-up. a) The solid line represents patients with biochemical non-response (*N*=258); the dotted line represent patients with a biochemical response (*N*=2,611). b) The solid line represents patients with an APRI>0.54 (*N*=1,022); the dotted line represents patients with an APRI≤0.54 (*N*=2,117). c) The gray dotted line line represent patients with high APRI and biochemical non-response (*N*=124); the black dotted line represents patients with high APRI and biochemical response (*N*=399); the black solid line represents patients with low APRI and biochemical non-response (*N*=2,012); the black dotted line represents patients with low APRI and biochemical response (*N*=604).

Major Hepatic Complications in Ursodeoxycholic Acid-Treated Patients With Primary Biliary Cholangitis: Risk Factors and Time Trends in Incidence and Outcome

Maren H. Harms, MD[1], Willem J. Lammers, MD, PhD[1], Douglas Thorburn, MD, PhD[2,3], Christophe Corpechot, MD, PhD[4], Pietro Invernizzi, MD, PhD[5], Harry L.A. Janssen, MD, PhD[6], Pier M. Battezzati, MD, PhD[7], Frederik Nevens, MD, PhD[8], Keith D. Lindor, MD, PhD[9,10], Annarosa Floreani, MD, PhD[11], Cyriel Y. Ponsioen, MD, PhD[12], Marlyn J. Mayo, MD, PhD[13], George N. Dalekos, MD, PhD[14], Tony Bruns, MD, PhD[15], Albert Parés, MD, PhD[16], Andrew L. Mason, MD, PhD[17], Xavier Verhelst, MD, PhD[18], Kris V. Kowdley, MD, PhD[19], Jorn C. Goet, MD[1], Gideon M. Hirschfield, MD, PhD[20], Bettina E. Hansen, MSc, PhD[1,6] and Henk R. van Buuren, MD, PhD[1] on behalf of the Global PBC Study Group

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VOLUME 113 | FEBRUARY 2018 www.nature.com/ajg

Table 2. Covariates associated with future development[a] of hepatic complications after 12 months of UDCA therapy

	Univariate analysis			Multivariable analysis		
	HR	95% CI	P	HR	95% CI	P
Age (per 10 years)	1.08	0.97–1.21	0.176	—	—	—
Age <40 years	0.74	0.50–1.10	0.140	—	—	—
Male sex	1.50	1.03–2.19	0.035	—	—	—
AMA negative	1.26	0.79–1.99	0.335	—	—	—
Year of diagnosis (per 10)	0.55	0.45–0.66	<0.001	0.73	0.60–0.89	0.002
Interval diagnosis—UDCA >2 years[a]	2.01	1.55–2.61	<0.001	—	—	—
Advanced disease[b]	4.34	3.18–5.92	<0.001	—	—	—
Total bilirubin ×ULN[c]	3.70	3.11–4.41	<0.001	—	—	—
ALP ×ULN[c]	2.37	1.94–2.90	<0.001	—	—	—
AST ×ULN[c]	3.26	2.73–3.91	<0.001	—	—	—
ALT ×ULN[c]	2.07	1.74–2.46	<0.001	—	—	—
Albumin ×LLN	0.01	0.004–0.04	<0.001	—	—	—
Platelets (per 50×10^3/mm^3)	0.47	0.41–0.54	<0.001	—	—	—
AST/ALT ratio[c]	1.62	1.20–2.19	0.002	—	—	—
APRI[d], continuous[c]	2.65	2.59–2.75	<0.001	—	—	—
APRI > 0.54	6.97	5.11–9.50	<0.001	5.32	3.82–7.41	<0.001
UK-PBC score (per 20)[e]	1.53	1.40–1.63	<0.001	—	—	—
UK-PBC score ≤median[e]	2.76	2.14–3.69	<0.001			
GLOBE-score, continuous	3.03	2.60–3.52	<0.001	—	—	—
Biochemical non-response[f]	5.52	4.17–7.33	<0.001	2.68	1.99–3.62	<0.001

Primary Biliary Cirrhosis and Cancer Risk: A Systematic Review and Meta-analysis

Yan Liang,* Zaixing Yang,* and Renqian Zhong

(Hepatology 2012;56:1409-1417)

Table 3. Pooled Relative Risks of Various Malignancies

Malignancy	No. of Studies	References	No. of Patients	Pooled RR or SIR (95% CI)	Heterogeneity	
					I^2 (%)	*P* Value
Overall cancer	9	11,12,21-27	6766	1.55 (1.28-1.83)	43.6	0.077
HCC	12	12,13,20-25,28-30	13,576	18.80 (10.81-26.79)	79.1	0.000
Breast cancer	9	1,11,12,21,23-27	5945	0.90 (0.58-1.23)	16.1	0.299
Kidney cancer	5	1,12,21,25,26	3221	2.06 (−1.36-5.48)	0	0.895
Colon cancer	5	11-13,21,26	8466	1.10 (0.81-1.40)	0	0.945
Lung cancer	3	21,24,27	3880	1.10 (0.43-1.76)	0	0.559
Colorectal cancer	3	1,25,27	739	1.13 (−0.26-2.52)	0	0.778
Rectal cancer	2	13,21	7838	1.00 (0.56-1.43)	0	0.910
Esophageal cancer	2	13,21	7838	1.29 (0.86-1.73)	0	0.851
Uterine cancer	2	21,25	2679	0.71 (−0.70-2.13)	0	0.613
Cervical cancer	2	11,26	420	3.81 (−4.85-12.47)	0	0.612
Prostate cancer	2	21,25	2679	0.27 (−1.25-1.79)	14.7	0.279
Bladder cancer	2	11,21	2315	1.64 (−1.07-4.35)	0	0.550
Thyroid cancer	2	12,25	767	4.13 (−6.07-14.34)	0	0.546
Skin melanoma	2	1,25	668	1.96 (−1.83-5.74)	0	0.400
Skin nonmelanoma	2	25,27	630	4.51 (−3.01-12.03)	0	0.550
Hodgkin disease	2	11,31	2387	4.86 (−3.00-12.71)	0	0.589
Non-Hodgkin lymphoma	3	1,25,31	2860	1.15 (0.36-1.94)	0	0.882

Stratification of hepatocellular carcinoma risk in primary biliary cirrhosis: a multicentre international study

Palak J Trivedi,[1,2] Willem J Lammers,[3] Henk R van Buuren,[3] Albert Parés,[4] Annarosa Floreani,[5] Harry L A Janssen,[3,6] Pietro Invernizzi,[7] Pier Maria Battezzati,[8] Cyriel Y Ponsioen,[9] Christophe Corpechot,[10] Raoul Poupon,[10] Marlyn J Mayo,[11] Andrew K Burroughs,[12,†] Frederik Nevens,[13] Andrew L Mason,[14] Kris V Kowdley,[15,16] Ana Lleo,[7] Llorenç Caballeria,[4] Keith D Lindor,[17,18] Bettina E Hansen,[3] Gideon M Hirschfield,[1,2] On behalf of the Global PBC Study Group

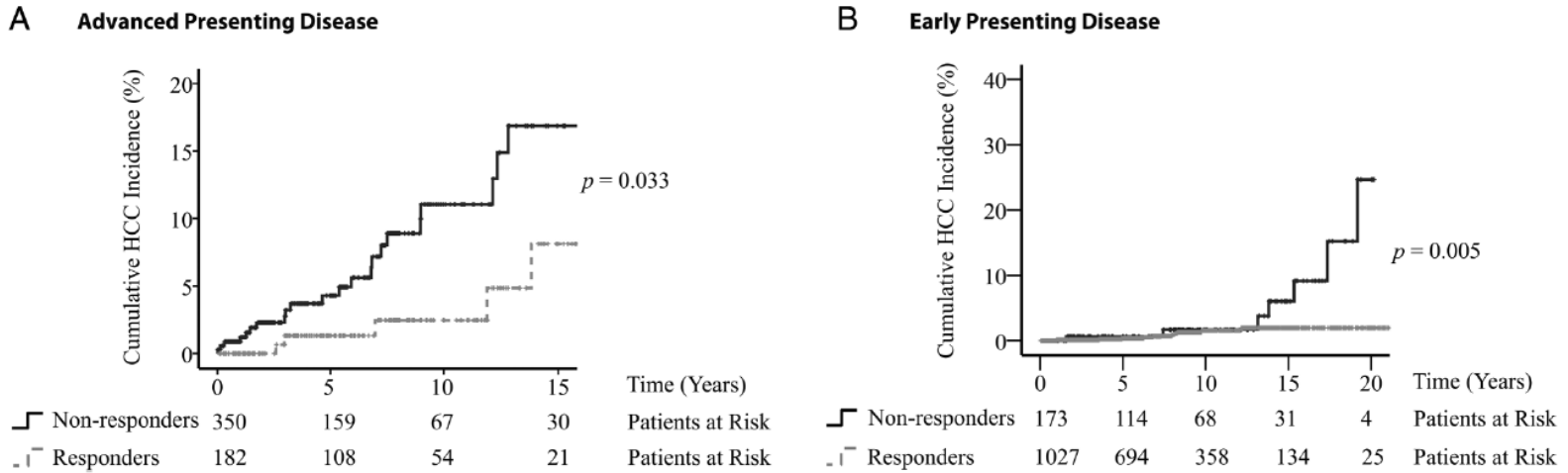


Figure 5 Hepatocellular carcinoma incidence in patients with varying disease stage stratified according to biochemical response. Kaplan-Meier estimate restricted to those with: (A) advanced presenting disease and biochemical non-response versus response, 11.2 vs 4.4 cases per 1000 patient-years; and (B) early presenting disease and biochemical non-response versus response, 4.7 vs 1.2 cases per 1000 patient-years. Analysis conducted in ursodeoxycholic acid-treated patients only in whom 12-month biochemical data were available to calculate response. Time measured in years following calculation of biochemical response.

Trivedi PJ, *et al*. *Gut* 2016;**65**:321–329. doi:10.1136/gutjnl-2014-308351

PBC: Transplant indications remain the same

- Elevated bilirubin
- Liver failure
- Cancer
- Intractable Itch

Gut, 1979, **20**, 137-140

Serum bilirubin: a prognostic factor in primary biliary cirrhosis

J. M. SHAPIRO, H. SMITH, AND F. SCHAFFNER

From the Division of Liver Diseases of the Department of Medicine and The Department of Bio-Statistics, Mount Sinai School of Medicine of The City University of New York, New York, USA

SUMMARY We followed up 55 patients with proven primary biliary cirrhosis for several years or until death. A graph of the level of serum bilirubin versus time that was constructed for each patient shows an initial stable period of variable length in which the serum bilirubin level remained constant. This was followed by a period of rapid rise in serum bilirubin which culminated in the patient's death. Whenever two successive serum bilirubin values taken six months apart exceeded 34 μmol/l (2·0 mg/dl) the patient had entered a late phase of disease and lived an average of 49 months. Ninety-five per cent confidence limits on survival time were 32-74 months. If two successive six month bilirubin values exceeded 102 μmol/l (6·0 mg/dl), calculated survival time was 25 months, and if two successive six month bilirubin values exceeded 170 μmol/l (10·0 mg/dl), survival time was 17 months. Fifteen of the 41 living patients had two consecutive serum bilirubin levels greater than 34 μmol/l (2·0 mg/dl). However, the slope of the rising bilirubin in the living patients is only 25 μmol/l/yr (1·5 mg/dl/yr) compared with 42 μmol/l/yr (2·5 mg/dl/yr) in the dead patients. This means that patients with this disease now may be living considerably longer.

Recommendations

42. EASL recommends considering patients for transplant assessment when they present with complications of cirrhosis, markers of disease severity (*e.g.* persistent elevated bilirubin values [50 μmol/L or 3 mg/dl] or MELD >15), or severe medically resistant pruritus. EASL recommends that listing for transplantation should follow local (usually national) guidelines (**II-2**, **1**).

43. EASL suggests that in patients with proven or likely recurrent PBC post liver transplant, the use of UDCA is safe and can improve liver biochemistry (**II-2**, **2**).

PBC CLINIC EASL GUIDELINES

"EASL Clinical Practice Guidelines: The Diagnosis and management of patients with Primary Biliary Cholangitis"
In lay terms: what does it mean for the patient?

pbc foundation
celebrating 20 years
Support for Life

QUESTION FOR YOUR DOCTOR:
Do I have a bilirubin level of over 50 μmol/L or 3 mg/dl? If so I wish to be referred to a liver transplant unit immediately.

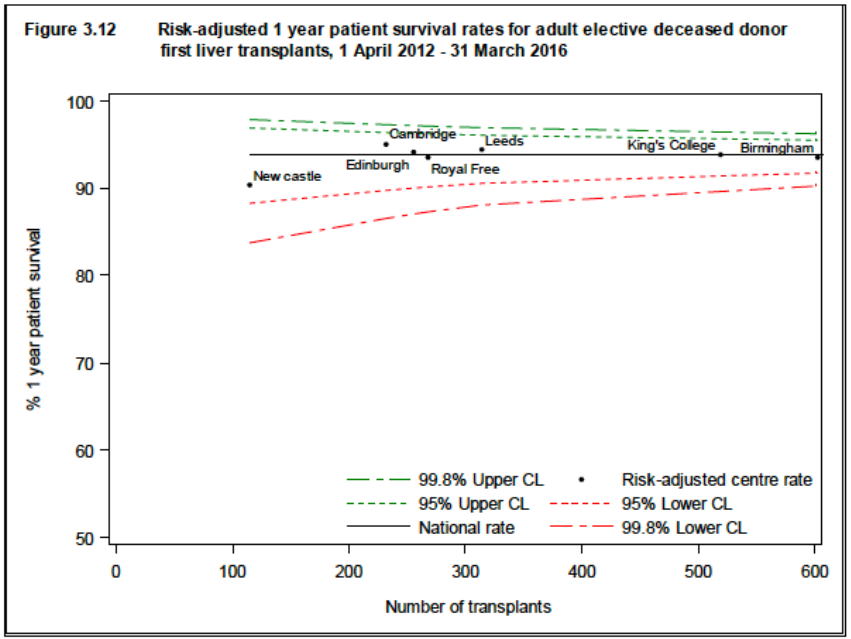
Clinical Practice Guidelines

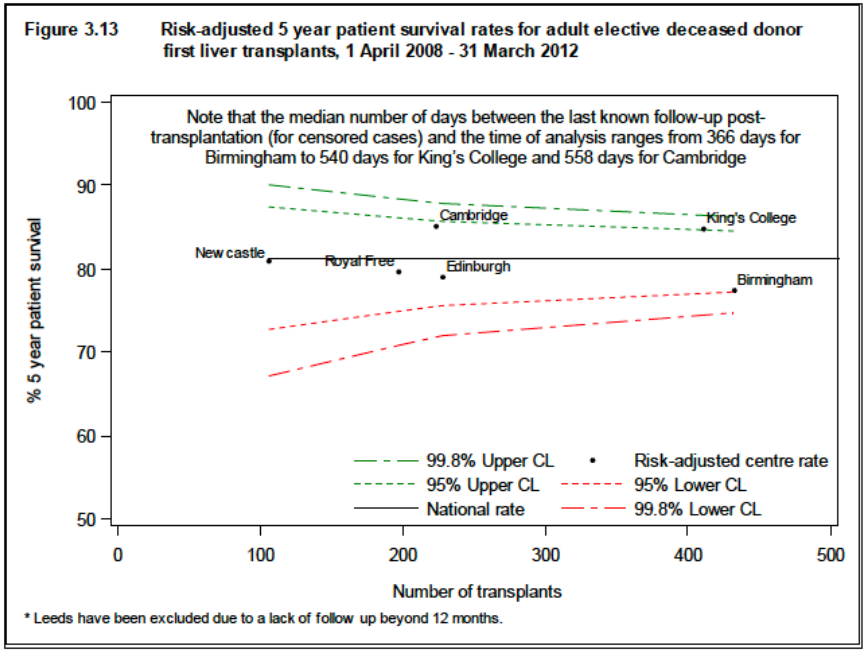
EASL JOURNAL OF HEPATOLOGY

EASL Clinical Practice Guidelines: The diagnosis and management of patients with primary biliary cholangitis☆

European Association for the Study of the Liver*

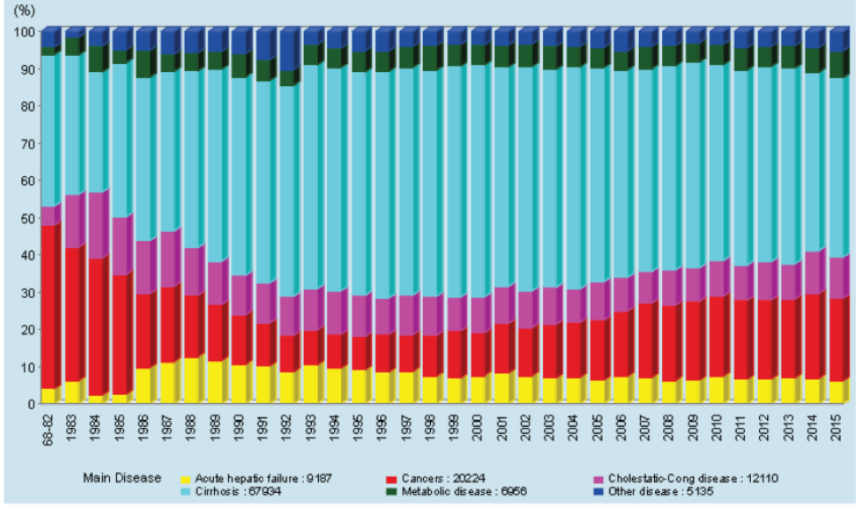
Post transplant survival is excellent!





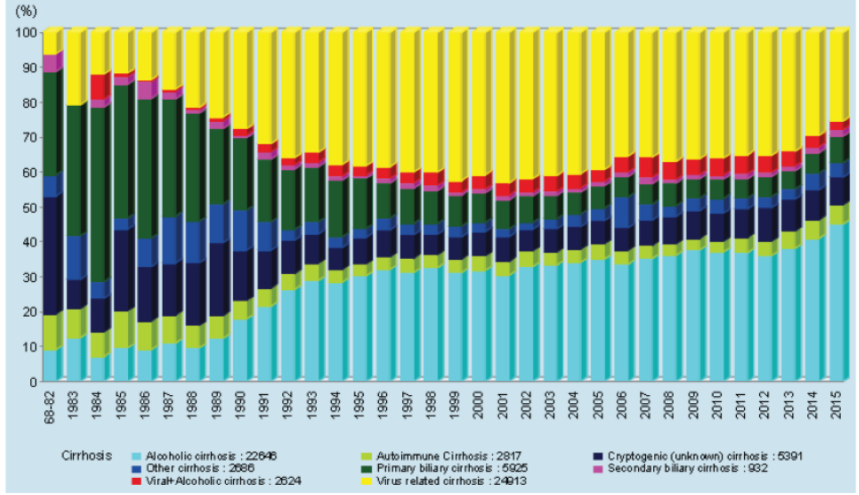
Evolution of Primary Disease Leading to Liver Transplantation in Europe

N = 121,546



Evolution of Cirrhosis leading to Liver Transplantation in Europe

N = 67,934



Evolving Frequency and Outcomes of Liver Transplantation Based on Etiology of Liver Disease

Ashwani K. Singal,[1,5] Praveen Guturu,[2] Bashar Hmoud,[3] Yong-Fang Kuo,[4] Habeeb Salameh,[3] and Russell H. Wiesner[1]

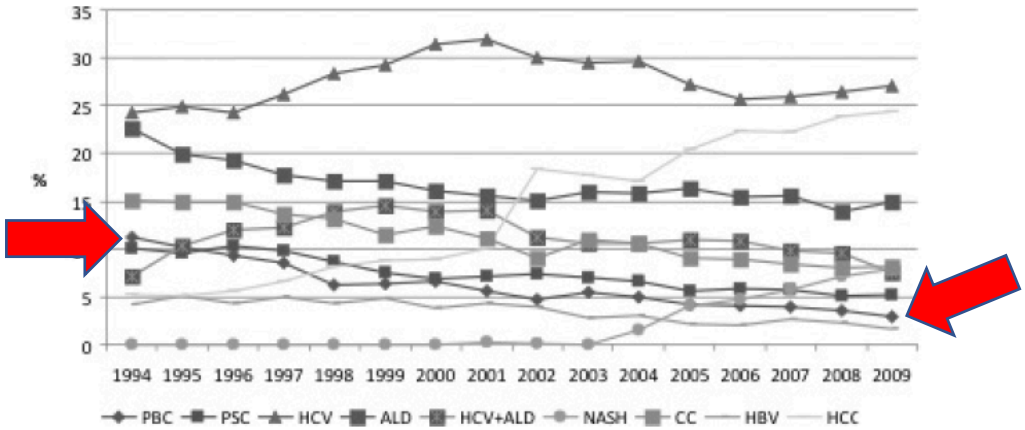
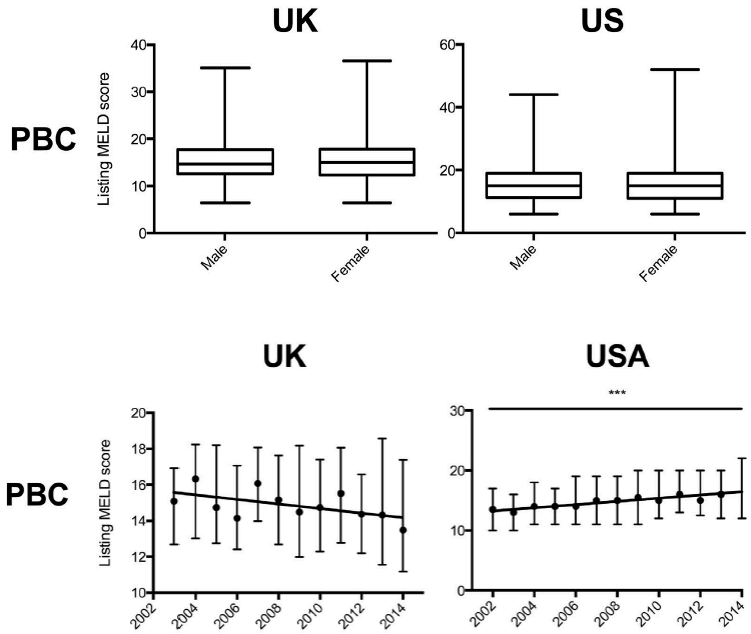


FIGURE 1. Proportion of patients transplanted annually during 1994–2009 for various indications.

Twenty-Year Comparative Analysis of Patients With Autoimmune Liver Diseases on Transplant Waitlists

Gwilym James Webb,[*,‡] Abbas Rana,[§] James Hodson,[‖] Mohammed Zeeshan Akhtar,[¶] James Walter Ferguson,[‡] James Max Neuberger,[‡] John Moore Vierling,[§] and Gideon Morris Hirschfield[*,‡]



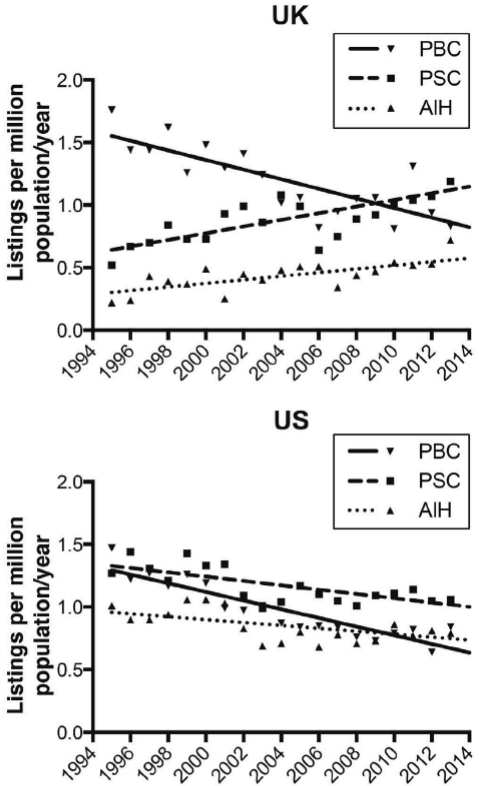


Figure 1. Adjusted listings for orthotopic liver transplantation over time. Listings for liver transplantation by year adjusted to population changes over the period studied for PBC, PSC, and AIH. In the United States, adjustments also were made for health insurance coverage. Super-urgent (in the United Kingdom) or status 1 (in the United States) listings, multi-region listings, patients listed for second or subsequent transplants, and patients listed for living-related liver donation were excluded. Linear regression suggested significant gradients over time for all conditions ($P < .01$ for all).

J Gastroenterol (2014) 49:324–331
DOI 10.1007/s00535-013-0782-5

The Japanese Society of Gastroenterology

ORIGINAL ARTICLE—LIVER, PANCREAS, AND BILIARY TRACT

Waiting list mortality of patients with primary biliary cirrhosis in the Japanese transplant allocation system

Takuya Genda · Takafumi Ichida · Shotaro Sakisaka · Michio Sata · Eiji Tanaka · Ayano Inui · Hiroto Egawa · Kouji Umeshita · Hiroyuki Furukawa · Seiji Kawasaki · Yukihiro Inomata

Table 2 Univariate and multivariate analysis of variables associated with waiting list mortality

Variables	Univariate			Multivariate		
	HR	95 % CI	*P* value	HR	95 % CI	*P* value
Age (per year of age)	1.04	1.03–1.05	<0.001	1.04	1.03–1.05	<0.001
Male gender	0.93	0.77–1.13	0.48			
Blood type						
A	1.00	Reference				
B	1.07	0.83–1.43	0.61			
O	1.13	0.90–1.43	0.29			
AB	1.26	0.90–1.77	0.17			
Etiology						
HCV	1.00	Reference				
BA	0.40	0.22–0.72	0.002			
PBC	1.62	1.21–2.16	0.001	1.79	1.34–2.39	<0.001
PSC	0.79	0.54–1.17	0.24			
HBV	0.77	0.56–1.05	0.10			
Alcohol	0.95	0.59–1.53	0.83			
AIH	0.77	0.34–1.74	0.52			
NASH	1.11	0.76–1.63	0.59			
HCC	1.46	1.05–2.05	0.003			
Metabolic disease	0.40	0.22–0.75	0.004			
Polycystic disease	0.26	0.10–0.70	0.008	0.27	0.10–0.73	0.01
Vascular disease	0.009	0.01–0.67	0.002			
Others	0.70	0.34–1.43	0.33			

AIH autoimmune hepatitis, *BA* biliary atresia, *HBV* hepatitis B virus, *HCC* hepatocellular carcinoma, *HCV* hepatitis C virus, *HR* hazard ratio, *NASH* non-alcoholic steatohepatitis, *PBC* primary biliary cirrhosis, *PSC* primary sclerosing cholangitis

Table 3 Comparison of patient characteristics between HCV and PBC

Variable	HCV ($n = 189$)	PBC ($n = 81$)	*P* value
Age (years)	55 (29–69)	52 (27–69)	0.02[a]
Gender (male/female)	143/46	15/66	<0.001[b]
Platelet count ($\times 10^4/\mu L$)	6.0 (1.7–49.0)	10.2 (2.2–42.3)	<0.001[a]
Albumin (g/dL)	2.8 (1.8–4.4)	2.8 (1.4–4.2)	0.96[a]
Total bilirubin (mg/dL)	2.7 (0.4–39.8)	7.2 (0.7–41.2)	<0.001[a]
Creatinine (mg/dL)	0.78 (0.4–7.4)	0.67 (0.37–2.83)	<0.001[a]
Prothrombin time (%)	54.7 (11.0–103.0)	62.2 (16.0–120.0)	0.001[a]
INR	1.51 (0.98–6.24)	1.32 (0.91–4.31)	0.001[a]
MELD score	15 (7–52)	17.5 (8–39)	0.002[a]
CTP score	10 (6–15)	10 (5–15)	0.27[a]
Medical point (1, 3/6, 9)	54/135	22/59	0.81[b]

Data are shown as median (range). Data were available for patients who were listed after June 22, 2006

CTP Child–Turcotte–Pugh, *HCV* hepatitis C virus, *INR* international normalized ratio, *MELD* model of end-stage liver disease, *PBC* primary biliary cirrhosis

[a] Mann–Whitney *U* test

[b] Chi-square test

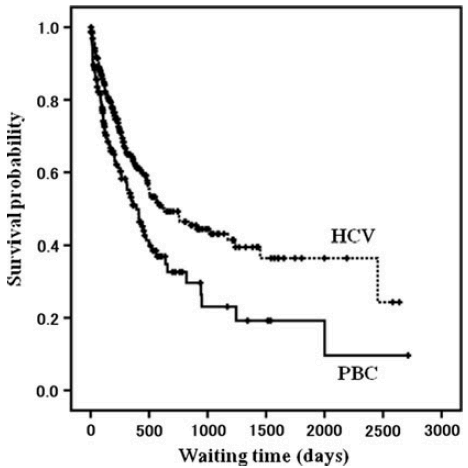


Fig. 2 Kaplan–Meier curves comparing the cumulative waiting list survival probability of patients with chronic hepatitis C (HCV, $n = 254$) and primary biliary cirrhosis (PBC, $n = 156$)

ORIGINAL ARTICLE

Primary biliary cirrhosis has high wait-list mortality among patients listed for liver transplantation

Ashwani K. Singal[1], Xiao Fang[2], Mohamed Kaif[1], Mohsen Hasanin[3,*], Brendan M. Mcguire[1], Yong-Fang Kuo[2] & Russell H. Wiesner[4]

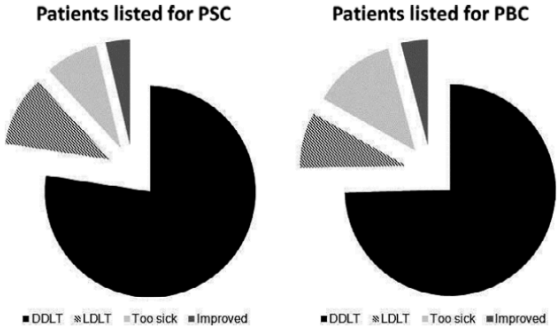


Table 3. Causes of death on wait-list mortality: comparison of patients listed for PBC or PSC.

	PSC (*N* = 418)	PBC (*N* = 575)
Cardiovascular *N* (%)	20 (4.8)	47 (8.2)
Organ failure *N* (%)	119 (28.5)	156 (27.1)
Infection *N* (%)	66 (15.8)	87 (15.1)
Hemorrhage *N* (%)	21 (5)	23 (4)
Other *N* (%)	34 (8.1)	49 (8.5)
Unknown *N* (%)	158 (37.8)	213 (37.1)

PSC, primary sclerosing cholangitis; PBC, primary biliary cirrhosis.

Table 2. Fine and Gray regression model for predictors of wait-list mortality.

Variable	Hazard ratio*	95% Confidence interval	*P*
Listing diagnosis: PBC versus PSC	1.25	1.07–1.47	0.006
Listing year	0.96	0.94–0.98	<.001
Age at listing in years	1.03	1.02–1.04	<0.001
Female versus male gender	1.31	1.12–1.54	0.001
Whites versus nonwhite race	1.00	0.85–1.17	0.95
BMI in kg/m^2 at listing	1.00	0.99–1.01	0.17
Diabetes versus no diabetes	1.07	0.90–1.29	0.44
Blood group B or AB versus A or O	0.74	0.61–0.91	0.003
MELD score at listing	1.02	1.01–1.03	<0.001
Serum albumin at listing	0.63	0.56–0.70	<0.001
UNOS regions			
5	Reference group		
1	0.89	0.64–1.23	0.48
2	0.81	0.64–1.03	0.09
3	0.32	0.24–0.44	<.001
4	1.00	0.80–1.24	0.97
6	0.49	0.33–0.73	<0.001
7	0.52	0.39–0.68	<0.001
8	0.81	0.63–1.04	0.09
9	0.95	0.74–1.23	0.71
10	0.53	0.41–0.70	<0.001
11	0.78	0.60–1.02	0.07
10 years increase in age	1.35	1.26–1.44	<0.001

PBC, primary biliary cirrhosis; PSC, primary sclerosing cholangitis; MELD, model for end-stage liver disease; BMI, body mass index.

*It is subdistribution hazard ratio as calculated from the Fine and Gray regression model.

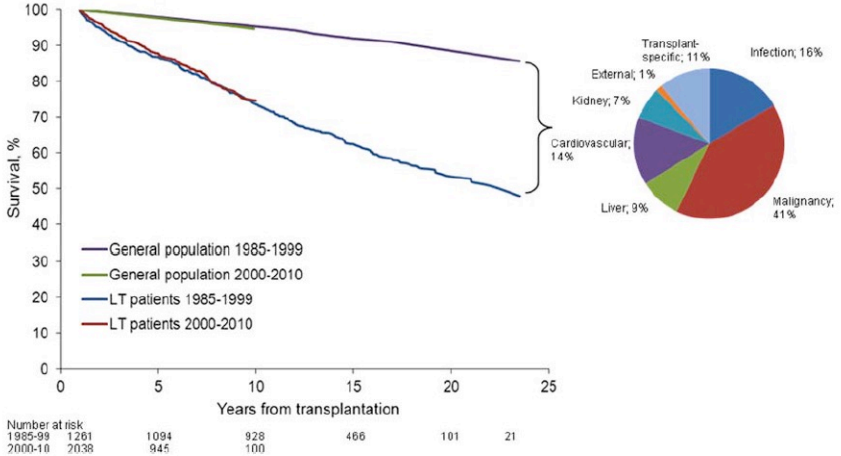


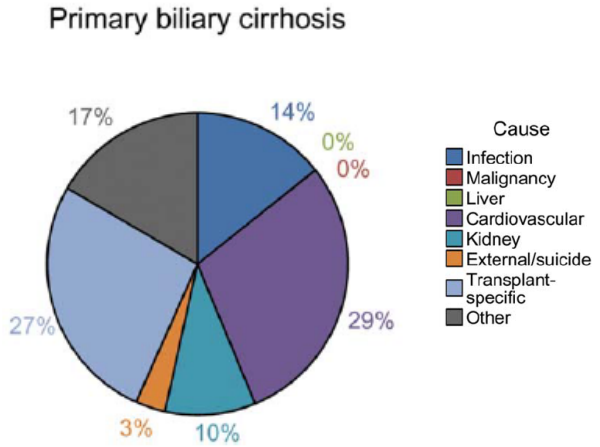
Table 3. Number of Deaths, Mortality Rate for Death Before Age 75, and Corresponding Standardized Mortality Rates (SMRs) With 95% Confidence Intervals (CIs) Compared to the General Population, According to Era of Transplantation and in Patient Subgroups

	Deaths	Mortality before age 75 per 100 000 follow-up years	SMR 1985-2010	95% CI	SMR 1985-1999	95% CI	SMR 2000-2010	95% CI
Male	441	3382	**7.6**	6.7-8.5	**7.8**	6.8-8.9	**8.6**	7.0-10.5
Female	322	2505	**5.0**	4.5-5.5	**5.2**	4.6-5.9	**5**	4.3-5.8
Age at transplantation								
0-14	43	1273	**83.4**	59.8-113.1	**76**	51.3-108.4	**94.4**	50.2-161.4
15-29	44	1574	**55.2**	39.8-74.6	**44.6**	29.9-64.1	**73**	40.9-120.4
30-44	128	2198	**21.6**	17.9-25.9	**20**	16.2-24.5	**23**	15.7-32.4
45-59	377	3545	**10.2**	9.1-11.3	**8.3**	7.3-9.3	**15**	12.5-17.9
60-	171	5245	**5.4**	4.5-6.3	**4**	3.2-4.9	**8.8**	7.0-10.8
Primary indication for transplantation								
Acute liver failure	61	2068	**6.4**	4.9-8.3	**5.9**	4.3-8.0	**7.6**	4.4-12.4
Alcoholic liver disease	102	4162	**17.7**	14.2-21.8	**8.3**	6.3-10.6	**24**	17.3-32.5
Hepatocellular carcinoma	84	8838	**32.6**	25.8-40.5	**38.4**	28.4-50.7	**18.8**	13.1-26.1
Primary biliary cirrhosis	92	2493	**3.3**	2.6-4.0	**4.9**	3.8-6.1	**2.9**	1.6-4.8
Primary sclerosing cholangitis	99	2462	**4.2**	3.4-5.2	**11**	8.3-14.2	**4.2**	3.0-5.7
Hepatitis C	88	6112	**12.4**	9.8-15.5	**23.1**	16.7-31.3	**9.2**	6.7-12.3
Any malignancy as the primary indication for transplantation								
Yes	114	7834	**20.02**	16.5-24.1				
No	649	2656	**5.15**	4.7-5.6				

Bold figures for significant levels.

Differences in Long-Term Survival Among Liver Transplant Recipients and the General Population: A Population-Based Nordic Study

Fredrik Åberg,[1,2] Mika Gissler,[3,4] Tom H. Karlsen,[5] Bo-Göran Ericzon,[6] Aksel Foss,[7] Allan Rasmussen,[8] William Bennet,[9] Michael Olausson,[9] Pål-Dag Line,[7] Arno Nordin,[1] Annika Bergquist,[10] Kirsten Muri Boberg,[5,11] Maria Castedal,[9] Christian Ross Pedersen,[8] and Helena Isoniemi[1]



Recurrent PBC: uncommon cause of graft loss or death

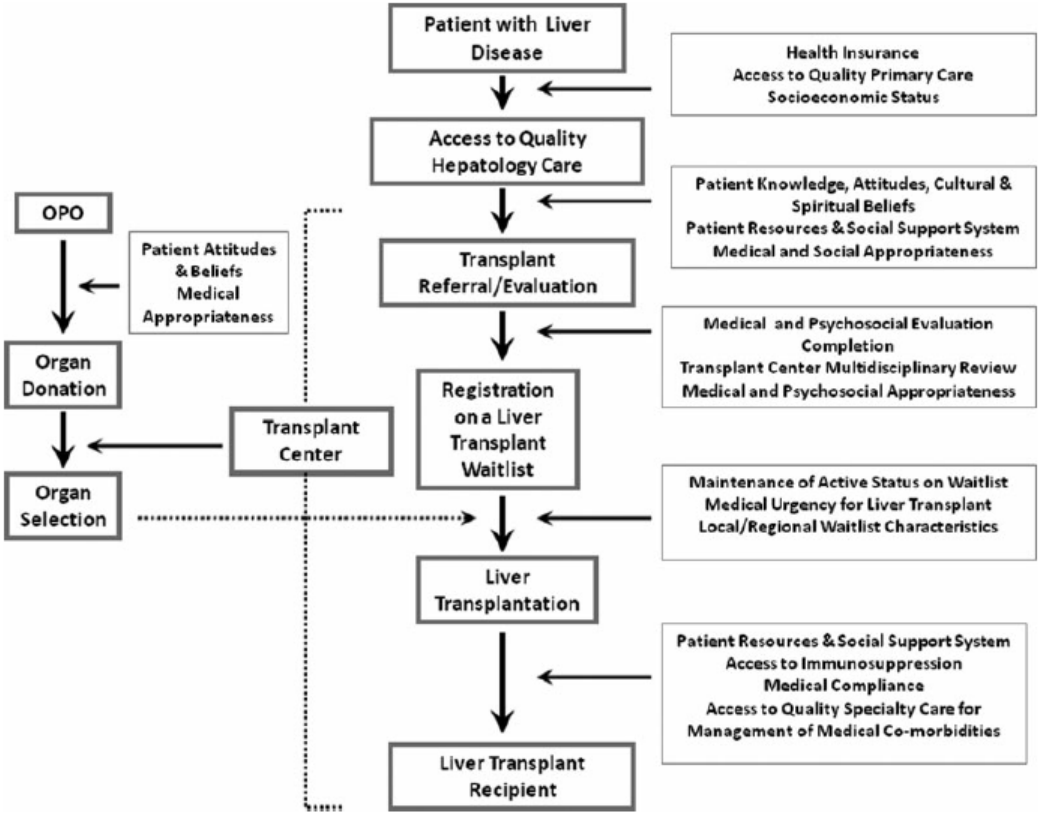
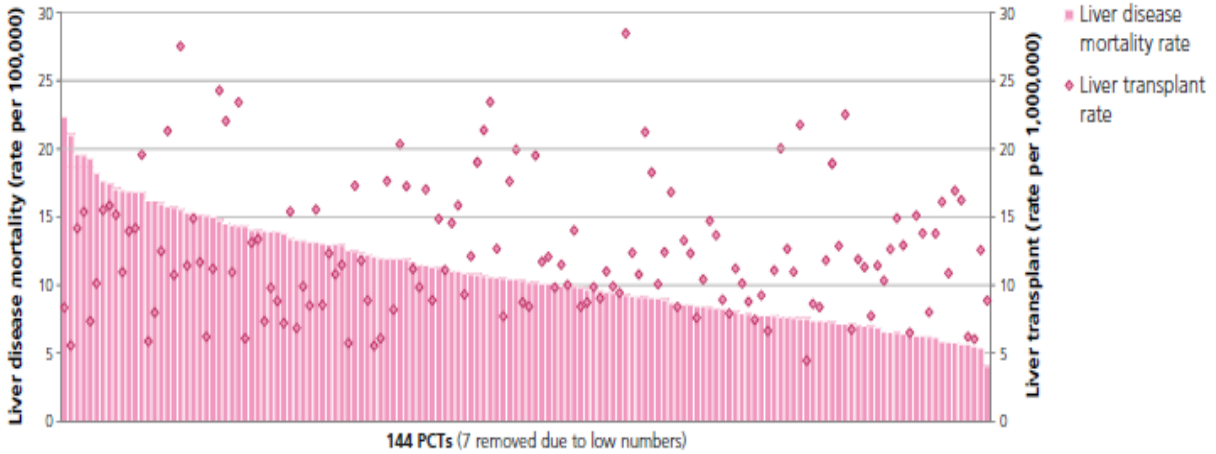


Figure 1: A conceptual model of disparities in the liver transplant process. The patient with end-stage liver disease must navigate through several complex steps in order to successfully undergo liver transplantation if indicated. Several factors affect the likelihood that a patient will be able to successfully complete each step in the process. This minireview aims to critique the available literature from each step in order to better understand racial/ethnic disparities in liver transplantation along a continuum of care.

Figure 6.1: Liver transplant rate pmp 2006/07–2010/11 (points) in relation to rate of chronic liver disease mortality (directly standardised) per 100,000 population aged under 75 years 2008–2010 (columns)



1 NHS Blood and Transplant. Liver Advisory Group. http://www.organdonation.nhs.uk/about_us/advisory_groups/lag/

2 NHS Blood and Transplant (2012) Organ Donation and Transplantation Activity Report 2011/12. http://www.organdonation.nhs.uk/statistics/transplant_activity_report/current_activity_reports/ukt/activity_report_2011_12.pdf

3 Data from seven PCTs have been removed due to low numbers (<5).

Transplantation and PBC.

- Urso improves outcomes in responders irrespective of disease stage.
- PBC reduced percentage of transplants; indications elevation of bilirubin, liver failure (? more common esp. men), intractable itch and cancer (poss. stratification; cirrhosis, non-response to Urso.).
- Possible increased mortality on waitlist
- Recurrence post transplant probably of little long term consequence
- Remember not only PBC cases “lost in the wilderness”, consider transplant in all cirrhosis UKELD >49 or decompensation episode.