Transplant Free Survival in HIV Associated Non Cirrhotic Portal Hypertension (NCPH)

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Background

- In the last decade there have been numerous reports of patients with HIV presenting with non-cirrhotic portal hypertension (NCPH). NCPH is associated with exposure to didanosine. We describe the natural history and clinical outcome in a large London cohort. .

Methods

- This is an retrospective observational cohort study. Demographics, laboratory, radiological and histological data were gathered on all patients at 2 London centres with non-cirrhotic portal hypertension.
- This was defined by portal hypertension, diagnosed clinically or on imaging and either a biopsy or elastography (score <11kPa) excluding cirrhosis.
- Patients with HBV SAg positive or HCV Ab positive were excluded.

Results

- 44 patients were identified who met our definition. Demographics are shown in Table 1, presenting features in Figure 1.
- Biopsy ws performed in 31/46 patients
- The most marked fibrosis reported was Ishak F3, majority mild or no fibrosis. No patient was cirrhotic.

Characteristic		N = 44 N(%)/Median (IQR)
Gender	M	26 (59%)
	F	18 (41%)
Ethnicity	Caucasian (Cauc)	20 (45%)
	Black African (BA)	24 (55%)
Age at presentation (years)		45.4 (40.6, 50.4)
History of DDI exposure		44/44 (100%)
Duration DDI exposure (months)		60 (54, 96)
Portal Vein Thrombosis		27/44 (63%)
Time since HIV diagnosis (years)		23 (20, 27)
HIV viral load <50 copies/ml at presentation		40/44 (90%)
CD4 cells/ml at presentation		291 (188, 393)

Table 1: Demographic and HIV related parameters

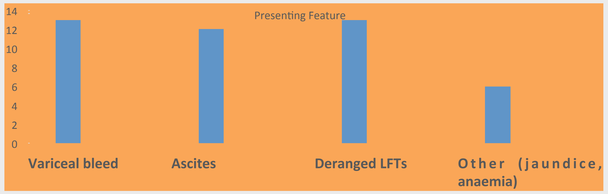
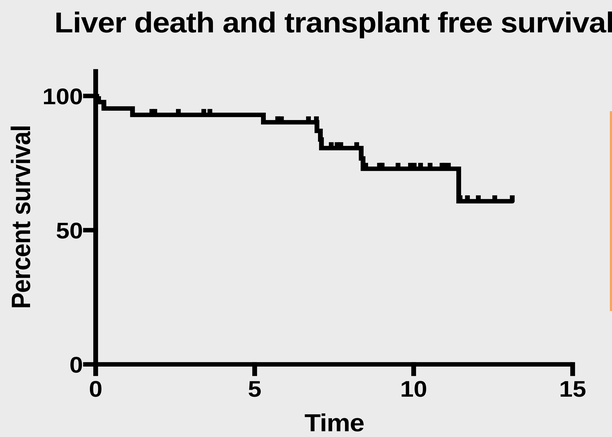
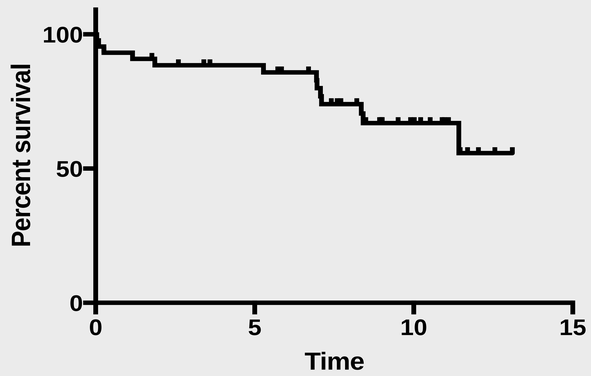


Figure 1: Presenting features of NCPH

Survival



Liver death and transplant free survival was 93% at 5 years, 72.9% at 10 years. All liver deaths were attributable to upper GI bleeding.



All cause death and transplant free survival was 88.4% at 5 years and 66.9% at 10 years

Abnormal coagulation

- 27 patients experienced chronic portal vein thrombosis, 17 had a full procoagulant screen prior to anticoagulation.
- Of those, 6/17 had low Protein S levels and 5/17 had low Protein C.
- 16/27 were anticoagulated with a total of 132 patient years of follow up.
- No major bleeding episodes occurred on warfarin

Liver Transplant

- 5 patients underwent orthotopic liver transplantation (OLT), see Table 2
- A further 2 patients underwent transplant assessment but were deemed too early for OLT at that time and are still in follow up. 2 more patients were listed for transplant but then lost to follow up.

Age	Gender (race)	CD4 (cells/ml)	Indication	UKELD	Warfarin pre-OLT	Years survival post OLT	Outcome
54	M (Cauc)	310	Encephalopathy	50	y	4.5	In Follow up
44	F (BA)	199	Diuretic resistant ascites	46	y	3 months	RIP. Complicated post OLT with thrombosis/ haemorrage /ARF Final event small and large bowel ischaemia secondary to SMV thrombus.
68	F (BA)	75	Encephalopathy plus diuretic resistant ascites	53	y	2.7	In Follow up
69	M (Cauc)	314	Encephalopathy post TIPSS	50	n	1.8	In Follow up
65	M (Cauc)	167	Synthetic failure	52	n	14 months	In Follow up

Table 2: OLT Indication and Outcome

Conclusions

We report on a large cohort of patients with a long duration of follow up for NCPH. We show that HIV associated NCPH is a serious condition with 27% of patients having died or undergone liver transplantation at 10 years. Anticoagulation was safe in this group. Many patients presented with subtle derangements of liver function and it is vital that HIV care providers maintain a high degree of clinical suspicion so that patients can receive joint care from both liver and HIV physicians.

Current Standardised Management NCPH

- Complete full chronic liver disease profile
- Fibroscan
- US liver for evidence portal hypertension
- Full anticoagulation screen (Protein C, S, AT III, JAK mutation, antiphospholipid, anticardiolipin, PNH screen)
- Liver biopsy
- Switch to least hepatotoxic ART
- Variceal screening
- Anticoagulation if PVT identified. Target INR 2-3