

CRF : The natural history of early porto-sinusoidal vascular disorder

Please collect the data in the following case report form (one per patient)

LOCAL INVESTIGATOR	
Last name :	
First name :	
Hospital :	
Country :	
Email address :	

Please send the following CRF and the data transfer agreement by email to :

Dr Edeline Kaze : kazeedeline@gmail.com

Demographic data, histological data and medical history

General information	
Participant code	
Referral hospital	
Age at diagnosis (years)	
Sex	Male YES/NO
	Female YES/NO
Height (cm)	
Weight (kg)	
Ethnicity	Caucasian YES/NO
	African YES/NO
	Asian YES/NO
	Latin America YES/NO
Date of PSVD diagnosis	
Date of liver biopsy (DD/MM/YYYY)	___ / ___ / ____
Histologic inclusion criteria	
Obliterative portal venopathy	YES/NO
Nodular regenerative hyperplasia	YES/NO
Incomplete septal fibrosis	YES/NO
Coexistence of 2 PSVD lesions	YES/NO If yes, describe the 2 coexisting lesions _____
Histologic exclusion criteria	
Biopsy size (only for core biopsy) < 20 mm	YES/NO
Histological portal tract changes	Portal vein obliteration YES/NO
	Portal fibrosis YES/NO
	Phlebosclerosis YES/NO
Other exclusion criteria	
Cirrhosis	YES/NO
Portal vein thrombosis	YES/NO
Budd-Chiari syndrome / hepatic venous outflow obstruction	YES/NO
Vascular invasion by a tumor	YES/NO
Severe comorbidity with limited estimated life expectancy	YES/NO (If yes, describe the severe comorbidity) _____
Presence at diagnosis of signs of PH Or	Gastric varices YES/NO
	Esophageal varices YES/NO

History of signs of PH	Ectopic varices YES/NO	
	Obvious porto-systemic collaterals at imaging YES/NO	
	Portal hypertensive bleeding YES/NO	
	Clinical ascites YES/NO	
	Platelet count < 150'000/mm3 YES/NO	
Spleen size ≥13 cm in the largest axis YES/NO		
Other histological data		
Indication for liver tissue	Abnormal liver tests YES/NO	
	Incidental diagnosis of PSVD YES/NO If yes, complete	Liver resection for metastasis YES/NO If yes describe the primary tumor _____
		Protocol biopsy after liver transplantation YES/NO
	Other (describe) _____	
Other (describe) _____		
Length of the sample (only for core biopsy) (mm)	YES /NOT AVAILABLE	
Fragmentation	YES/NO	
Number of portal tracts (only for core biopsy)	_____/NOT AVAILABLE	
Valoration of biopsy quality	Adequate / Not adequate	
Architectural changes	YES/NO	
Sinusoidal dilatation	YES/NO	
Sinusoidal dilatation grade	0 YES/NO	
	I Slight YES/NO	
	II moderate YES/NO	
	III several YES/NO	
Slight perisinusoidal fibrosis	YES/NO	
Dilated vessels like a cavernoma	YES/NO	
METAVIR score	F0 YES/NO	
	F1 YES/NO	
	F2 YES/NO	
Lobular inflammation score	0 YES/NO	
	1 YES/NO	
	2 YES/NO	
Portal inflammation score	0 YES/NO	
	1 YES/NO	
	2 YES/NO	
	3 YES/NO	
Conditions associated with PSVD at diagnosis		
History of drug/Toxine exposure	YES/NO If yes, complete	
	Chemical exposure YES/NO	Exposure start date (DD/MM/YYYY) ____/____/_____

	If yes, complete	Exposure end date (DD/MM/YYYY) ____/____/____
		Copper sulphate YES/NO
		Vinyl chloride monomer YES/NO
		Thorium sulphate YES/NO
		Spanish toxic oil YES/NO
	Vitamine exposure YES/NO If yes, complete	Arsenic as Fowler's solution YES/NO
		Vitamin A supplements YES/NO
		Exposure start date (DD/MM/YYYY) ____/____/____
	Drug exposure YES/NO If yes, complete	Exposure end date (DD/MM/YYYY) ____/____/____
		Exposure start date (DD/MM/YYYY) ____/____/____
		Drug indication (describe) _____
		Stavudine YES/NO
		Didanosine YES/NO
		Azathioprine YES/NO
		6-Mercaptopurine YES/NO
		6-Thioguanine YES/NO
		Oxaliplatin YES/NO
		Methotrexate YES/NO
		Busulfan YES/NO
		Doxorubicin YES/NO
		Cyclophosphamide YES/NO
		Chlorambucil YES/NO
		Bleomycin YES/NO
	Other YES/NO If yes, complete	Cytarabine YES/NO
		Carmustine YES/NO
		Cytosine arabinoside YES/NO
		Exposure start date (DD/MM/YYYY) ____/____/____
History of immunological disorders	Exposure end date (DD/MM/YYYY) ____/____/____	
	Indication (describe) _____	
History of immunological disorders	YES/NO If yes, complete	
	Date of diagnosis (DD/MM/YYYY) ____/____/____	
	Chronic glomerulonephritis YES/NO if yes specify the subtype _____	
	Common variable immune deficiency YES/NO	
	Autoimmune hepatitis YES/NO	
	Systemic lupus erythematosus YES/NO	

	Systemic sclerosis (scleroderma) YES/NO
	Rheumatoid arthritis YES/NO
	HIV YES/NO
	Celiac disease YES/NO
	POEMS syndrome YES/NO
	Autoimmune thyroiditis YES/NO
	Multiple sclerosis YES/NO
	Felty's syndrome YES/NO
	Polyarteritis nodosa YES/NO
	Antiphospholipid syndrom YES/NO
	Crohn's disease YES/NO
	Ulcerative colitis YES/NO
History of hemocoagulative disorders	YES/NO
	If yes, complete
	Date of diagnosis (DD/MM/YYYY) ____ / ____ / _____
	Aplastic anaemia YES/NO
	Idiopathic thrombocytopenic purpura YES/NO
	Sickle cell anemia YES/NO
	Myeloproliferative disorders YES/NO
	If yes complete
	Polycythemia vera YES/NO
	Essential thrombocythemia YES/NO
	Primary myelofibrosis YES/NO
	Chronic myeloid leukemia YES/NO
	Hodgkin's lymphoma YES/NO
	Non-Hodgkin's lymphoma YES/NO
	Chronic lymphocytic leukemia YES/NO
	Multiple myeloma YES/NO
	Macroglobulinemia YES/NO
	Protein C deficiency YES/NO/NOT SCREENED
	Protein S deficiency YES/NO/NOT SCREENED
	Prothrombin (Factor II) gene mutation YES/NO/NOT SCREENED
	Factor V Leiden YES/NO/NOT SCREENED
	Antiphospholipid syndrome YES/NO/NOT SCREENED
	ADAMTS13 deficiency YES/NO/NOT SCREENED
	MTHFR deficiency YES/NO/NOT SCREENED
History of congenital and Hereditary Disorders	YES/NO
	If yes, complete
	Date of diagnosis (DD/MM/YYYY) ____ / ____ / _____
	Portal vein agenesis YES/NO
	Congenital heart defect YES/NO, if yes describe _____
	Turner's syndrome YES/NO
	Adams-Oliver syndrome YES/NO
	TERT mutations YES/NO/NOT SCREENED
	Cystic fbrosis YES/NO
	Familial cases YES/NO
	KCNN3 mutation YES/NO
History of infection	More than one abdominal infection at birth (including omphalitis) YES/NO
	Tuberculosis YES/NO
History of congestive heart failure	YES/NO

Other comorbidities at diagnosis

Other comorbidities at diagnosis	YES/NO If yes, describe and add date of comorbidity diagnosis DD/MM/YYYY ____ / ____ / ____	
Other causes of chronic liver disease	Active Alcohol consumption YES/NO (If yes ____ unit/day) and number of consumption years ____ years)	
	History of alcohol consumption YES/NO If yes , complete	alcohol consumption start date (DD/MM/YYYY) ____ / ____ / ____
		alcohol consumption end date (DD/MM/YYYY) ____ / ____ / ____
		quantity (unit/day) ____ /day
	Hepatitis B virus	HBsAg Positive YES/NO/Not tested
		Antiviral therapy YES/NO If yes which agent (describe) _____
	Hepatitis C virus	YES/NO/Not tested
		Sustained virological response YES/NO
		If yes, date (DD/MM/YYYY) ____ / ____ / ____
	Autoimmune YES/NO (If yes describe) _____	
	Other etiology YES/NO (If yes describe) _____	
History of liver transplantation	YES/NO If yes, complete	
	Date of liver transplantation (DD/MM/YYYY) : ____ / ____ / ____	
	Immunossuppressive regimen at PSVD diagnosis (describe) : _____	

Data at diagnosis

Thrombophilia screen performed at diagnosis	
YES/NO	
If yes, complete	
Date of thrombophilia screen	DD/MM/YYYY ____ / ____ / ____
Antithrombin deficiency	YES/NO/NOT SCREENED Value (%): _____ Normal range: (80-140%)
Protein C deficiency	YES/NO/NOT SCREENED Value (%): _____ Normal range: (80-140%)
Protein S deficiency	YES/NO/NOT SCREENED Value (%): _____ Normal range: (80-140%)
Activated protein C resistance	YES/NO/NOT SCREENED
Factor V mutation	YES/NO/NOT SCREENED
Factor II mutation	YES/NO/NOT SCREENED
Antiphospholipid syndrome	YES/NO/NOT SCREENED
Lupus anticoagulant antibodies IgM	YES/NO/NOT SCREENED
Lupus anticoagulant antibodies IgG	YES/NO/NOT SCREENED
Anticardiolipin antibodies IgM	YES/NO/NOT SCREENED
Anticardiolipin antibodies IgG	YES/NO/NOT SCREENED
Anti-beta2-GP I antibodies IgM	YES/NO/NOT SCREENED
Anti-beta2-GP I antibodies IgG	YES/NO/NOT SCREENED
Myeloproliferative disorder	YES/NO/NOT SCREENED if yes, describe
	Polycythemia vera YES/NO
	Essential thrombocythemia YES/NO
	Primary myelofibrosis YES/NO
Marrow bone biopsy	YES/NO If yes, complete
	Chronic myeloid leukemia YES/NO
	date (DD/MM/YYYY) ____ / ____ / ____
	Results (describe) _____
JAK 2 mutation	YES/NO/NOT SCREENED
CALR mutation	YES/NO/NOT SCREENED
Paroxysmal nocturnal hemoglobinuria	YES/NO/NOT SCREENED
Ongoing oral contraception at diagnosis	YES/NO if yes, which agent (describe) _____
Laboratory data at diagnosis	
Date	DD/MM/YYYY ____ / ____ / ____
Bilirubine total/direct/indirect (mg/dl)	
Aspartate aminotransferase (AST) (U/L)	

Alanine aminotransferase (ALT) (U/L)	
Alkaline phosphatase (U/L)	
Gamma-glutamyl transpeptidase (GGT) (U/L)	
Albumine (g/L)	
Hemoglobine (g/L).	
Hematocrite (%)	
Leucocytes(x10 ⁹ /L)	
Plateletes (x10 ⁹ /L)	
INR. Protrombine t (%)	
Creatinine (mg/dL)	
MELD score	
Child Score (number)	
Imaging at diagnosis	
Date of 1st liver imaging (the closer imaging test to diagnosis)	DD/MM/YYYY ____ / ____ / ____
Select the type of imaging study, select more than one if more than one.	US / CT-Scan / MRI
Hepatic artery diameter (mm)	YES /NO/NOT AVAILABLE
Hypertrophy of segment IV	YES /NO/NOT AVAILABLE
Hypertrophy of segment I	YES /NO/NOT AVAILABLE
Nodular liver surface	YES /NO/NOT AVAILABLE
FibroScan® (Echosens, Paris, France) at diagnosis YES / NOT DONE If yes, complete	
Date	DD/MM/YYYY ____ / ____ / ____
Performer	Experienced hepatology nurse/Hepatologist/Other (describe) _____
Liver stiffness (kPa)	YES /NOT AVAILABLE
IQR (kPa)	YES /NOT AVAILABLE
Spleen stiffness (kPa)	YES /NOT AVAILABLE
IQR (kPa)	YES /NOT AVAILABLE
**If several FibroScan® please add all of them with the date	

Data during follow-up

Last clinical information		
Extra-hepatic comorbidities occurring follow-up and different from those at diagnosis	YES/NO If yes , complete	Date of diagnosis (DD/MM/YYYY) ____/____/____
		Describe_____
Endoscopic data	YES/NO If yes, complete	
	Date : DD/MM/YYYY	____/____/____
	Esophageal varices (specify :small/medium/large) YES /NO/NOT AVAILABLE	
	Gastric varices (specify : IGV I – IGV II) YES /NO/NOT AVAILABLE	
	Esophagogastric varices (specify : GOV I – GOV II) YES /NO/NOT AVAILABLE	
	Portal hypertensive gastropathy Specify : Slight/moderate/severe YES /NO/NOT AVAILABLE	
	Ectopic varices (describe the location) YES /NO/NOT AVAILABLE	
Ascites	YES/NO/NOT AVAILABLE If yes, complete	
	Date of occurrence (DD/MM/YYYY) ____/____/____	
	Specify : grade I –II – III	
	Diuretic controlled YES/NO/NOT AVAILABLE	
	Refractory ascites YES/NO/NOT AVAILABLE	
	Paracentesis YES/NO/NOT AVAILABLE. How many per month ____/month	
Spontaneous bacterial peritonitis	YES/NO/NOT AVAILABLE If yes : date (DD/MM/YYYY) ____/____/____	
Acute kidney injury	YES/NO/NOT AVAILABLE If yes, date (DD/MM/YYYY) ____/____/____ specify: grade IA, IB, II, III	
Hepatorenal syndrome	YES/NO If yes, date (DD/MM/YYYY) ____/____/____ specify : Type I/II	
Hepatic encephalopathy	YES/NO/NOT AVAILABLE If yes , complete	
	Date of occurrence (DD/MM/YYYY) ____/____/____	
	Specify grade I – II – III – IV	
	Pharmacological control lactulose	

	YES/NO/NOT AVAILABLE	
	Rifaximine needed to control YES/NO/NOT AVAILABLE	
	Chronic HE: YES/NO/NOT AVAILABLE. Number of episodes Maximum severity I-II-III-IV	
	Precipitant YES/NO/NOT AVAILABLE. If yes which one (constipation, infection. ..describe) _____	
Portal hypertension hemorrhage	YES/NO/NOT AVAILABLE If yes , complete	
	Date of occurrence (DD/MM/YYYY) ____/____/____	
	Gastric varices YES/NO/NOT AVAILABLE (Specify : IGV I – IGV II)	
	Esophagogastric varices YES/NO/NOT AVAILABLE (Specify : GOV I – GOV II)	
	Ectopic varices YES/NO/NOT AVAILABLE Describe the location _____	
	Portal hypertensive gastropathy YES/NO/NOT AVAILABLE Slight/moderate/severe	
	Hepatopulmonar Syndrome	
Portal cholangiopathy	YES/NO/NOT AVAILABLE If yes, date (DD/MM/YYYY) ____/____/____	
	Severity I, II, III	
	Ursodeoxycholic acid treatment YES/NO If yes, date (DD/MM/YYYY) ____/____/____	
Hepatocellular carcinoma	YES/NO/NOT AVAILABLE If yes, complete	
	date of diagnosis (DD/MM/YYYY) ____/____/____	
	Within Milan Criteria YES/NO/NOT AVAILABLE	
Nodules	YES/NO If yes,complete	Date of nodule diagnosis (DD/MM/YYYY) ____/____/____
		Number of nodules 1 / 2-4 / >5
	Nodules growth	YES/NO/NOT AVAILABLE If yes,complete
		date of growth (DD/MM/YYYY) ____/____/____

	Nodules biopsy	YES/NO/NOT AVAILABLE If yes, complete
		Date of biopsy (DD/MM/YYYY) ____/____/____
		describe histology _____
Last laboratory data		
Date	DD/MM/YYYY ____/____/____	
Bilirubine total/direct/indirect (mg/dl)		
Aspartate aminotransferase (AST) (U/L)		
Alanine aminotransferase (ALT) (U/L)		
Alkaline phosphatase (U/L)		
Gamma-glutamyl transpeptidase (GGT) (U/L)		
Albumine (g/L)		
Hemoglobine (g/L).		
Hematocrite (%)		
Leucocytes(x10 ⁹ /L)		
Plateletes (x10 ⁹ /L)		
INR. Protrombine t (%)		
Creatinine (mg/dL)		
MELD score		
Child Score (number)		
Last imaging		
Date	DD/MM/YYYY ____/____/____	
If absence of thrombosis during follow-up = date and findings of the last liver imaging		
If presence of thrombosis during follow-up = date and findings of liver imaging showing thrombosis		
Select the type of imaging study, select more than one if more than one.	US / CT-Scan / MRI	
Hepatic artery diameter (mm)	YES /NO/NOT AVAILABLE	
Hypertrophy of segment IV	YES /NO/NOT AVAILABLE	
Hypertrophy of segment I	YES /NO/NOT AVAILABLE	
Nodular liver surface	YES /NO/NOT AVAILABLE	
Splenomegaly Spleen size in the largest axis (cm)	YES /NO/NOT AVAILABLE	
Portal velocity at diagnosis (cm/s)	YES /NO/NOT AVAILABLE	
Thrombosis of right or left portal branch	YES /NO/NOT AVAILABLE	
Thrombosis of portal trunk	YES /NO/NOT AVAILABLE	
Thrombosis of splenic vein	YES /NO/NOT AVAILABLE	
Thrombosis of mesenteric vein	YES /NO/NOT AVAILABLE	
Obvious porto-systemic collaterals	YES /NO/NOT AVAILABLE	
Ascites	YES /NO/NOT AVAILABLE	
Other findings and the techniques at which was found	Describe _____	

FibroScan® (Echosens, Paris, France) during follow up YES / NOT DONE If yes, complete	
Date	DD/MM/YYYY ____ / ____ / ____
Performer	Experienced hepatology nurse/Hepatologist/Other (describe) _____
Liver stiffness (kPa)	YES /NOT AVAILABLE
IQR (kPa)	YES /NOT AVAILABLE
Spleen stiffness (kPa)	YES /NOT AVAILABLE
IQR (kPa)	YES /NOT AVAILABLE
**If several FibroScan® please add all of them with the date	
Hepatic and cardiovascular hemodynamic at follow-up YES / NOT DONE If yes, complete	
Date of hepatic vein catheterization	DD/MM/YYYY ____ / ____ / ____
Hepatic Venous Pressure Gradient (mmHg)	
Wedged hepatic vein pressure (mmHg):	
Free hepatic vein pressure (mmHg):	
Medical intervention during follow-up	
Transjugular intrahepatic portosystemic shunt (TIPS)	YES/NO If yes, complete
	date (DD/MM/YYYY) ____ / ____ / ____
	Indication for TIPS (describe) _____
Liver transplantation	YES/NO If yes, complete
	Date (DD/MM/YYYY) ____ / ____ / ____
	Indication for liver transplantation (describe) _____
Tumors occurring during follow-up	
Extrahepatic tumors	YES/NO If yes, complete
	Diagnosis date (DD/MM/YYYY) ____ / ____ / ____
	Type of tumor (describe) _____

Mortality	
Liver-related mortality	YES/NO
	If yes, complete
	Date (DD/MM/YYYY) ____ / ____ / ____ cause of death (describe) _____
Non-liver-related mortality	YES/NO
	If yes, complete
	Date (DD/MM/YYYY) ____ / ____ / ____ cause of death (describe) _____
Last follow-up	
Date of last follow up	DD/MM/YYYY ____ / ____ / ____
Patient lost to follow-up	YES/NO