

Data Set Name: analysis.sas7bdat

Num	Variable	Type	Len	Format	Informat	Label
1	id	Num	8			
2	YMHS1C2	Char	5	\$5.	\$5.	Is the research subject listed for liver transplantation? (2=Yes)
3	Age_at_baseline	Num	8			Age at Baseline
4	ms_Race	Char	5			race
5	YMHA1B2	Char	5	\$5.	\$5.	gender (1=male, 2=female)
6	YMHS1C13A	Char	2	\$2.	\$2.	Polysplenia syndrome (1=yes)
7	YMHS1E1H	Char	2	\$2.	\$2.	Medication received: Prophylactic antibiotics for cholangitis (1=Yes)
8	YMHS1E1B	Char	2	\$2.	\$2.	Medication received: Vitamin D (1=Yes)
9	YMHS1E1G	Char	2	\$2.	\$2.	Medication received: Ursodeoxycholic acid (1=Yes)
10	YMHS1E1L	Char	2	\$2.	\$2.	Medication received: non-selective beta blockers (Propranalol or nadalol) (1=Yes)
11	YANOC10A	Char	3	\$3.	\$3.	Gastrointestinal malformations: Abdominal heterotaxia (1=Yes)
12	YANOC10B	Char	3	\$3.	\$3.	Gastrointestinal malformations: Abnormal pancreatic structure (1=Yes)
13	YANOC10C	Char	3	\$3.	\$3.	Gastrointestinal malformations: Absent hepatic artery (1=Yes)
14	YANOC10D	Char	3	\$3.	\$3.	Gastrointestinal malformations: Absent portal vein (1=Yes)
15	YANOC10E	Char	3	\$3.	\$3.	Gastrointestinal malformations: Annular pancreas (1=Yes)
16	YANOC10F	Char	3	\$3.	\$3.	Gastrointestinal malformations: Anomalous hepatic artery (1=Yes)
17	YANOC10G	Char	3	\$3.	\$3.	Gastrointestinal malformations: Anomalous portal vein (1=Yes)
18	YANOC10H	Char	3	\$3.	\$3.	Gastrointestinal malformations: Duodenal/Jejunal atresia (1=Yes)
19	YANOC10I	Char	3	\$3.	\$3.	Gastrointestinal malformations: Esophageal atresia (1=Yes)
20	YANOC10J	Char	3	\$3.	\$3.	Gastrointestinal malformations: Gastroschisis (1=Yes)
21	YANOC10K	Char	3	\$3.	\$3.	Gastrointestinal malformations: Imperforated anus (1=Yes)
22	YANOC10L	Char	3	\$3.	\$3.	Gastrointestinal malformations: Intestinal malrotation (1=Yes)
23	YANOC10M	Char	3	\$3.	\$3.	Gastrointestinal malformations: Micrognathia/Agnathia (1=Yes)
24	YANOC10N	Char	3	\$3.	\$3.	Gastrointestinal malformations: Midline liver (1=Yes)
25	YANOC10O	Char	3	\$3.	\$3.	Gastrointestinal malformations: Omphalocele (1=Yes)
26	YANOC10P	Char	3	\$3.	\$3.	Gastrointestinal malformations: Pancreatic cyst (1=Yes)
27	YANOC10Q	Char	3	\$3.	\$3.	Gastrointestinal malformations: Preduodenal portal vein (1=Yes)
28	YANOC10R	Char	3	\$3.	\$3.	Gastrointestinal malformations: Right-sided stomach (1=Yes)
29	YANOC10S	Char	3	\$3.	\$3.	Gastrointestinal malformations: Tracheoesophageal Fistula (TEF) (1=Yes)
30	YANOC10T	Char	3	\$3.	\$3.	Gastrointestinal malformations: Transverse (left-sided) liver (1=Yes)
31	YANOC10U	Char	3	\$3.	\$3.	Gastrointestinal malformations: Other (1=Yes)
32	YMHS2DDE	Char	20	\$20.	\$20.	Past 12 month Cholangitis Events
33	YMHS2IIS	Char	300	\$300.	\$300.	Bone Fracture Site
34	age_at_consent	Num	8			Age at consent (years)
35	age_at_kasai	Num	8			age at Kasai/HPE (days)
36	age_at_now	Num	8			Current age at evaluation (years)
37	cardiac_hx	Num	8			Has Congenital Heart Disease? (1=Yes, 0=No)

Num	Variable	Type	Len	Format	Informat	Label
38	yphec4	Num	8			Right mid arm circumference (cm)
39	yphec5	Num	8			Right triceps skinfold thickness (mm)
40	yphec6	Num	8			Subscapular skinfold thickness (mm)
41	WAZ	Num	8			Weight z-scores
42	HAZ	Num	8			Height z-scores
43	tbili_flag	Num	8			TB level normal (≤ 1.5 mg/dl) (1=Yes, 0=No)
44	AST_flag	Num	8			AST level normal (≤ 45 IU/L) (1=Yes, 0=No)
45	ALT_flag	Num	8			AST level normal (≤ 40 IU/L) (1=Yes, 0=No)
46	albumin_flag	Num	8			Serum Alb level level normal (≥ 40 3.3 g/dl) (1=Yes, 0=No)
47	GGTP_flag	Num	8			GGT level normal (≤ 55 U/L) (1=Yes, 0=No)
48	INR_flag	Num	8			INR normal (≤ 1.3) (1=Yes, 0=No)
49	platelet_flag	Num	8			Plate count ($\geq 150 \times 10^3/\text{mm}^3$) (1=Yes, 0=No)
50	haz_flag	Num	8			Age-adjusted height z-score > -2 (1=Yes, 0=No)
51	waz_flag	Num	8			Age-adjusted weight z-score > -2 (1=Yes, 0=No)
52	choly_prophy_flag	Num	8			Absence of Need for Cholangitis Prophylaxis (1=Yes, 0=No)
53	ascites_flag	Num	8			Absence of ascites (1=Yes, 0=No)
54	hps_flag	Num	8			Absence of Hepatoplummonary syndrome (1=Yes, 0=No)
55	cholangitis_flag	Num	8			Experience cholangitis at least once since HPE (1=good, 0=no)
56	cholangitis_flag_12	Num	8			Cholangitis occurred within the past 12 months (1=good, 0=no)
57	evbleed_flag	Num	8			Absence of Variceal bleeding (1=Yes, 0=No)
58	fracture_flag	Num	8			Absence of Fractures (1=Yes, 0=No)
59	fracture_flag_12	Num	8			Fractures occurred within the past 12 months (1=good, 0=no)
60	nspleensize	Num	8			Spleen size (cm)
61	chocolate	Num	8			King's College ideal survivor (1=yes, 0=no)
62	all_labs	Num	8	3.		Total # Subjects with ALL above Lab Indices Normal (1=Yes, 0=No)
63	all_complications	Num	8	3.		Absence of Known Complications of Chronic Liver Disease (1=yes, 0=no)
64	ideal_survivor	Num	8			>5 -year native liver survivor with ideal outcomes (1=Yes, 0=No)
65	PedsQL	Num	8			PedsQL Total Score ≥ 69.7 (normal) (1=Yes, 0=No)
66	GI_hx	Num	8			Has Gastrointestinal malformations? (1=Yes, 0=No)
67	vitamin	Num	8			Medication received: Vitamin supplement (1=Yes)
68	diuretics	Num	8			Medication received: Diuretics (1=Yes)
69	yilbb1mg	Num	8			Total bilirubin (mg)
70	yilbb2mg	Num	8			Indirect bilirubin (mg)
71	yilbb3mg	Num	8			Direct bilirubin (mg)
72	yilbb4mg	Num	8			Unconjugated bilirubin (mg)
73	yilbb5mg	Num	8			Conjugated bilirubin (mg)
74	yilbb6ul	Num	8			AST (ul)
75	yilbb7ul	Num	8			ALT (ul)
76	yilbb9gl	Num	8			Albumin (gl)

Num	Variable	Type	Len	Format	Informat	Label
77	yilbb11ul	Num	8			GGTP (ul)
78	yilbb33pl	Num	8			Platelets
79	yilbb13in	Num	8			INR
80	YPHEC1KG	Num	8			Weight (kg)
81	YPHEC2CM	Num	8			Height (cm)
82	zyphec4	Num	8			Right MAC z-score
83	zyphec5	Num	8			Right triceps z-score
84	zyphec6	Num	8			Right subscap z-score

Data Set Name: ba_vh_18aug22.sas7bdat

Num	Variable	Type	Len	Format	Label
1	sex	Num	8	SEX.	Sex
2	LiverTxp	Num	8	YN.	Liver transplant
3	ParticipantID	Char	25	\$25.	De-Identified Subject ID
4	race	Num	8	RACEFMT.	Race
5	KasaiAge	Num	8		Age at HPE (days)
6	WeightKg	Num	8		Weight (kg)
7	HeightCm	Num	8		Height (cm)
8	SpleenSize	Num	8	BEST12.	Spleen size (cm below costal margin)
9	ASTUnitsL	Num	8	BEST12.	AST (U/L)
10	ALTUnitsL	Num	8	BEST12.	ALT (U/L)
11	TotalBilirubinMgdl	Num	8		Total bilirubin (mg/dl)
12	GGTPUnitsL	Num	8	BEST12.	GGTP (U/L)
13	INR	Num	8	BEST12.	INR
14	PlateletsCnt	Num	8	BEST12.	Platelet count (10 ³ /mm ³)
15	AlkPhosphUnitsL	Num	8	BEST12.	Alkaline phosphatase (U/L)
16	AlbuminGdl	Num	8		Albumin (g/dl)
17	sHPE	Num	8	YN.	Functioning HPE
18	waz	Num	8		Weight z-score
19	haz	Num	8		Height z-score
20	whpct	Num	8		Weight-for-height percentile
21	bmicat	Num	8	BMICATF.	BMI
22	mhx	Num	8	YN.	VH reported on medical history form
23	bleed	Num	8	YN.	VH during follow-up
24	bleedhx	Num	8	YN.	History of VH prior to enrollment
25	PELD	Num	8	BEST12.	PELD at transplant
26	age_bsl_y	Num	8		Age at baseline visit (years)
27	age_bsl_m	Num	8		Age at baseline visit (months)
28	age_txp_y	Num	8		Age at transplant (years)
29	TxpDeathEnd	Num	8	YN.	Transplant/death outcome
30	time_to_TxpDeath	Num	8		Time to transplant/death (years)
31	BleedEnd	Num	8	BLEEDENDF.	VH outcome
32	time_to_Bleed	Num	8		Time to first bleed (years)
33	Txp6w	Num	8	YN.	Transplant within 6 weeks of bleed
34	Death6w	Num	8	YN.	Death within 6 weeks of bleed
35	TxpDeath6w	Num	8	YN.	Transplant or death within 6 weeks of bleed
36	age_bleed_y	Num	8		Age at first bleed (years)
37	spleenplate	Num	8	SPLEENPLATEF.	Thrombocytopenia/Splenomegaly

Num	Variable	Type	Len	Format	Label
38	spleenplate2	Num	8	SPLEENPLATE2F.	Thrombocytopenia/Splenomegaly
39	time_fu_y	Num	8		Follow-up time (years)
40	mhx_sens	Num	8	YN.	VH reported on medical history form [sensitivity analysis]
41	Bleed_sens	Num	8	YN.	VH during follow-up [sensitivity analysis]
42	BleedEnd_sens	Num	8	BLEEDENDF.	VH outcome [sensitivity analysis]
43	time_to_Bleed_sens	Num	8		Time to first bleed (years) [sensitivity analysis]
44	bmi	Num	8		BMI
45	bmiz	Num	8		BMI z-score
46	basic	Num	8	BASICF.	Study

Data Set Name: bileacids_niddk_21dec2022.sas7bdat

Num	Variable	Type	Len	Format	Informat	Label
1	study	Char	5			Study
2	TBiliM6	Num	8			Total bilirubin (mg/dl), 6 months
3	CBiliM6	Num	8			Conjugated bilirubin (mg/dl), 6 months
4	sex	Num	8	SEX.		Sex
5	Ethnicity	Num	8	HISPANICF.		Ethnicity
6	LiverTxp	Num	8	YN.		Liver transplant
7	ParticipantID	Char	25	\$25.		De-Identified Subject ID
8	RaceDSMB	Num	8	RACEFMTDSMB.		Race
9	TREATMENT	Char	15	\$15.	\$15.	START treatment arm
10	basM	Num	8	YN.		BASM
11	wazM6	Num	8			Weight z-score, 6 months
12	hazM6	Num	8			Height z-score, 6 months
13	SpleenSizeM6	Num	8	BEST12.		Spleen size (cm), 6 months
14	wazY2	Num	8			Weight z-score, year 2
15	hazY2	Num	8			Height z-score, year 2
16	SpleenSizeY2	Num	8	BEST12.		Spleen size (cm), year 2
17	BileAcidsM6	Num	8			Serum bile acids (umol/L), 6 months
18	UBiliM6	Num	8			Unconjugated bilirubin (mg/dl), 6 months
19	ASTM6	Num	8	BEST12.		AST (U/L), 6 months
20	ALTM6	Num	8	BEST12.		ALT (U/L), 6 months
21	GGTPM6	Num	8	BEST12.		GGTP (U/L), 6 months
22	PlateletsM6	Num	8	BEST12.		Platelet count, 6 months
23	NaM6	Num	8	BEST12.		Sodium (mmol/L), 6 months
24	INRM6	Num	8	BEST12.		INR, 6 months
25	AlbuminM6	Num	8			Albumin (g/dl), 6 months
26	VitDM6	Num	8			Vitamin D (D25OH) (ng/ml), 6 months
27	INRY2	Num	8	BEST12.		INR, year 2
28	AlbuminY2	Num	8			Albumin (g/dl), year 2
29	BileAcidsY2	Num	8			Serum bile acids (umol/L), year 2
30	CBiliY2	Num	8			Conjugated bilirubin (mg/dl), year 2
31	TBiliY2	Num	8			Total bilirubin (mg/dl), year 2
32	UBiliY2	Num	8			Unconjugated bilirubin (mg/dl), year 2
33	VitDY2	Num	8			Vitamin D (D25OH) (ng/ml), year 2
34	ALTY2	Num	8	BEST12.		ALT (U/L), year 2
35	ASTY2	Num	8	BEST12.		AST (U/L), year 2
36	GGTPY2	Num	8	BEST12.		GGTP (U/L), year 2
37	NaY2	Num	8	BEST12.		Sodium (mmol/L), year 2

Num	Variable	Type	Len	Format	Informat	Label
38	PlateletsY2	Num	8	BEST12.		Platelet count, year 2
39	CholangitisHx	Num	8	YN.		History of cholangitis
40	BleedHx	Num	8	YN.		History of GI bleed
41	FractureHx	Num	8	YN.		History of fracture
42	Ascites	Num	8	YN.		Ascites
43	Cholangitis	Num	8	YN.		Cholangitis
44	Bleed	Num	8	YN.		GI bleed
45	Fracture	Num	8	YN.		Fracture
46	Spleno	Num	8	YN.		Splenomegaly
47	Thrombo	Num	8	YN.		Thrombocytopenia
48	UrsoM6	Num	8	YN.		Ursodeoxycholic acid use, 6 months
49	UrsoY2	Num	8	YN.		Ursodeoxycholic acid use, year 2
50	TSBA_Measured	Num	8			Bile acids (umol/L), measured in sample
51	TCA	Num	8	15.2		TCA (umol/L)
52	TUDCA	Num	8	15.2		TUDCA (umol/L)
53	TCDC	Num	8	15.2		TCDC (umol/L)
54	TDCA	Num	8	15.2		TDCA (umol/L)
55	TLCA	Num	8	15.2		TLCA (umol/L)
56	GCA	Num	8	15.2		GCA (umol/L)
57	GUDCA	Num	8	15.2		GUDCA (umol/L)
58	GCDCA	Num	8	15.2		GCDCA (umol/L)
59	GDCA	Num	8	15.2		GDCA (umol/L)
60	GLCA	Num	8	15.2		GLCA (umol/L)
61	DCA	Num	8	15.2		DCA (umol/L)
62	LCA	Num	8	15.2		LCA (umol/L)
63	T_G	Num	8	BEST.		T/G (umol/L)
64	CA	Num	8	15.2		CA (umol/L)
65	CDCA	Num	8	15.2		CDCA (umol/L)
66	UDCA	Num	8	15.2		UDCA (umol/L)
67	Conj_CA	Num	8	15.2		Conj CA (umol/L)
68	Conj_CDCA	Num	8	15.2		Conj CDCA (umol/L)
69	Conj_UDCA	Num	8	15.2		Conj UDCA (umol/L)
70	Unconj_Conj_CA	Num	8	COMMA15.7		Unconj/Conj CA (umol/L)
71	Unconj_Conj_CDCA	Num	8	COMMA15.7		Unconj/Conj CDCA (umol/L)
72	Unconj_Conj_UDCA	Num	8	COMMA15.7		Unconj/Conj UDCA (umol/L)
73	TCA_GCA	Num	8	BEST.		TCA/GCA (umol/L)
74	TCDC_GCDCA	Num	8	BEST.		TCDC/GCDCA (umol/L)
75	TUDCA_GUDCA	Num	8	BEST.		TUDCA/GUDCA (umol/L)
76	age_hpe_d	Num	8			Age at KP (days)

Num	Variable	Type	Len	Format	Informat	Label
77	TxpDeath	Num	8	YN.		Transplant/death
78	BSL_TxpDeath_m	Num	8			Time from bile acids measurement to transplant/death (months)
79	HPE_TxpDeath_m	Num	8			Time from KP to transplant/death (months)
80	AscitesBSL	Num	8	YN.		Ascites at 6 months post-KP
81	SplenoBSL	Num	8	YN.		Splenomegaly at 6 months post-KP
82	ThromboBSL	Num	8	YN.		Thrombocytopenia at 6 months post-KP
83	CEPHBSL	Num	8	YN.		CEPH at 6 months post-KP
84	BSL_AscitesEnd_m	Num	8			Time from bile acids measurement to new onset ascites (months)
85	HPE_AscitesEnd_m	Num	8			Time from KP to new onset ascites (months)
86	AscitesEnd	Num	8	ENDF.		Ascites (endpoint)
87	BSL_SplenoEnd_m	Num	8			Time from bile acids measurement to new onset splenomegaly (months)
88	HPE_SplenoEnd_m	Num	8			Time from KP to new onset splenomegaly (months)
89	SplenoEnd	Num	8	ENDF.		Splenomegaly (endpoint)
90	BSL_ThromboEnd_m	Num	8			Time from bile acids measurement to new onset thrombocytopenia (months)
91	HPE_ThromboEnd_m	Num	8			Time from KP to new onset thrombocytopenia (months)
92	ThromboEnd	Num	8	ENDF.		Thrombocytopenia (endpoint)
93	BSL_CholangitisEnd_m	Num	8			Time from bile acids measurement to cholangitis (months)
94	HPE_CholangitisEnd_m	Num	8			Time from KP to cholangitis (months)
95	CholangitisEnd	Num	8	ENDF.		Cholangitis (endpoint)
96	BSL_BleedEnd_m	Num	8			Time from bile acids measurement to GI bleed (months)
97	HPE_BleedEnd_m	Num	8			Time from KP to GI bleed (months)
98	BleedEnd	Num	8	ENDF.		GI Bleed (endpoint)
99	BSL_FractureEnd_m	Num	8			Time from bile acids measurement to fracture (months)
100	HPE_FractureEnd_m	Num	8			Time from KP to fracture (months)
101	FractureEnd	Num	8	ENDF.		Fracture (endpoint)
102	CEPH	Num	8	YN.		CEPH
103	BSL_CEPHEnd_m	Num	8			Time from bile acids measurement to new onset CEPH (months)
104	HPE_CEPHEnd_m	Num	8			Time from KP to new onset CEPH (months)
105	CEPHEnd	Num	8	ENDF.		CEPH (endpoint)

Data Set Name: manuscript_data_25jun19.sas7bdat

Num	Variable	Type	Len	Format	Informat	Label
1	ChiLDReN_StudyID	Char	50	\$50.	\$50.	ChiLDReN_StudyID
2	sex	Num	8	SEX.	6.	Sex
3	LiverTxp	Num	8	YN.		Transplant
4	study	Char	5			Study
5	WeightFailure	Num	8	YN.		Weight growth failure
6	HeightFailure	Num	8	YN.		Height growth failure
7	INR	Num	8	8.1		INR
8	AlbuminGdl	Num	8			Albumin (g/dl)
9	TotalBilirubinMgdl	Num	8			Total bilirubin (mg/dl)
10	ALTUnitsL	Num	8	BEST12.		ALT (U/L)
11	ASTUnitsL	Num	8	BEST12.		AST (U/L)
12	GGTPUnitsL	Num	8	BEST12.		GGTP (U/L)
13	PlateletsCnt	Num	8	BEST12.		Platelet count
14	AscitesHx	Num	8	YN.		History of ascites
15	HPSHx	Num	8	YN.		History of HPS
16	SpleenHx	Num	8	YN.		History of splenomegaly
17	CholangitisHx	Num	8	YN.		History of cholangitis
18	BleedHx	Num	8	YN.		History of GI bleed
19	GIAnomaly	Num	8	YN.		Gastrointestinal anomaly
20	HeartAnomaly	Num	8	YN.		Cardiovascular anomaly
21	asplenia_polysplenia	Num	8	YN.		Asplenia/Polysplenia
22	Antibiotics	Num	8	YN.		Antibiotics
23	Urso	Num	8	YN.		Ursodeoxycholic acid
24	age_kasai_d	Num	8			Age at Kasai (days)
25	race	Num	8	RACEF.		Race
26	hispanic	Num	8	YN.		Hispanic ethnicity
27	TxpDeathEnd	Num	8	YN.		Transplant/death
28	time_to_TxpDeath	Num	8			Time to transplant/death (years)
29	ComboEnd	Num	8	COMBOF.		SE
30	AscitesFirst	Num	8	YN.		First SE was ascites
31	HPSFirst	Num	8	YN.		First SE was HPS
32	BleedFirst	Num	8	YN.		First SE was GI bleed
33	time_to_Combo	Num	8			Time to first sentinel event (years)
34	age_lastfu_y	Num	8			Age last observed (years)
35	age_consent_m	Num	8			Age at consent (months)
36	Death	Num	8	YN.		Death