

Data Set Name: a1atbaseline.sas7bdat

Num	Variable	Type	Len	Label
1	GROUP	Num	8	Eligibility Group
2	RACECAT	Char	1	Race category
3	DMGAB02	Char	3	Gender
4	DMGAB03	Char	3	Ethnicity
5	AST	Num	8	AST U/L
6	ALT	Num	8	ALT U/L
7	INR	Num	8	INR
8	GGTP	Num	8	GGTP U/L
9	WBC	Num	8	WBC 10 ³ /mm ³
10	AST_ALT_RATIO	Num	8	AST/ALT Ratio
11	PHT	Num	8	Portal Hypertension
12	PHT_US	Num	8	Portal Hypertension ultrasound finding
13	LIVER_DISEASE	Num	8	Liver disease severity: Mild vs. Severe
14	AGE	Num	8	Age (at consent)
15	IHX2B01_AGE	Num	8	Age first noticed symptoms of liver disease
16	IHX2B02_AGE	Num	8	Age of diagnosis of CLiC liver disease
17	ZZ	Num	8	ZZ Genotype/phenotype
18	SZ	Num	8	SZ Genotype/phenotype
19	PRETX	Num	8	Liver transplant
20	ID	Num	8	De-Identified Subject ID

Data Set Name: a1atlongv4.sas7bdat

Num	Variable	Type	Len	Format	Informat	Label
1	bmi	Num	8			BMI
2	WeightZScore	Num	8			Weight-for-age Z
3	headcz	Num	8			Head circ-for-age Z
4	BMIZScore	Num	8			BMI-for-age Z
5	HeightZScore	Num	8			Height-for-age Z
6	PiZZPheno	Num	8	YN.		PiZZ phenotype
7	PiSZPheno	Num	8	YN.		PiSZ phenotype
8	Gender	Num	8	SEX.	6.	Sex
9	Ethnicity	Num	8	ETHF.	6.	Ethnicity
10	FinalStatus	Num	8	ALLLEAVE.		Final status
11	SubjectID	Char	8			LOGIC subject ID
12	AnyLiverTxp	Num	8	YN.		Liver transplant
13	RTriSkin	Num	8	8.1		Right triceps skinfold thickness (mm)
14	RArmSizeCm	Num	8			Right mid arm circumference (cm)
15	INR	Num	8	8.1		INR
16	AlbuminGdl	Num	8			Albumin (g/dL)
17	ALTUnitsL	Num	8	BEST12.		ALT (U/L)
18	ASTUnitsL	Num	8	BEST12.		AST (U/L)
19	GGTPUnitsL	Num	8	BEST12.		GGTP (U/L)
20	PlateletsCnt	Num	8	BEST12.		Platelets (10 ³ /mm ³)
21	BiliTotalMgdl	Num	8			Total or (unconjugated + conjugated) bilirubin (mg/dL)
22	BiliDirectMgdl	Num	8			Direct or conjugated bilirubin (mg/dL)
23	AgeAtExam	Num	8			Age at visit (years)
24	Visit	Num	8	VISITFMT.		Visit
25	AgeAtTxp	Num	8			Age (years) at liver transplant
26	race	Num	8	RACECATF.		Race
27	PHT	Num	8	YN.		PHT status (on this visit)
28	PHTBaseline	Num	8	YN.		PHT on entry
29	TxpEnd	Num	8	YN.		Liver transplant/death
30	PHTEnd	Num	8	YN.		PHT
31	TxpEndYrs	Num	8			Time to liver transplant/death (years)
32	PHTEndYrs	Num	8			Time to PHT (years)
33	ExamYrs	Num	8			Time to visit (years)
34	NeoCholestasis	Num	8	YN.		Neonatal cholestasis
35	RespiratoryProblems	Num	8	YN.		Respiratory problems

Data Set Name: algsbl.sas7bdatt

Num	Variable	Type	Len	Format	Informat	Label
1	agemos	Num	8			Age at exam (months)
2	sex	Num	3	SEXFMT.		Sex
3	bmi	Num	8			BMI
4	WeightZScore	Num	8			weight-for-age Z
5	BMIZScore	Num	8			BMI-for-age Z
6	HeightZScore	Num	8			height-for-age Z
7	Ethnicity	Num	8	ETHNICITYFMT.	6.	Ethnicity
8	SubjectID	Char	8			LOGIC subject ID
9	Race	Num	8	RACEFMT.		Race
10	IHxKasai	Num	8	YNMFMT.		Kasai (initial history)
11	IHxSurgeryDrainAny	Num	8	YNMFMT.		Drainage procedure (initial history)
12	IHxALGSPPS	Num	8	YNMFMT.		Peripheral pulmonary stenosis
13	IHxALGSPulmStenosis	Num	8	YNMFMT.		Pulmonary valve stenosis
14	IHxALGSTetraFallot	Num	8	YNMFMT.		Tetralogy of Fallot
15	IHxALGSVentricDef	Num	8	YNMFMT.		Ventricular septal defect
16	IHxALGSAtriaDef	Num	8	YNMFMT.		Atrial septal defect
17	IHxALGSDeepEyes	Num	8	YNMFMT.		Deep-set eyes
18	IHxALGSForehead	Num	8	YNMFMT.		Broad forehead
19	IHxALGSChin	Num	8	YNMFMT.		Pointed chin
20	IHxALGSPostEmbrotaxon	Num	8	YNMFMT.		Posterior embryotoxon
21	IHxALGSAxenfeld	Num	8	YNMFMT.		Axenfeld's anomaly
22	IHxALGSButterfly	Num	8	YNMFMT.		Butterfly vertebrae
23	IHxALGSDyspKidney	Num	8	YNMFMT.		Dysplastic kidney
24	IHxALGSSingleKidney	Num	8	YNMFMT.		Single kidney
25	IHxALGSRTA	Num	8	YNMFMT.		Renal tubular acidosis
26	IHxALGSHeartOther	Num	8	YNMFMT.		Other cardiac anomaly
27	IHxALGSEyesOther	Num	8	YNMFMT.		Other eye condition
28	IHxALGSSkelOther	Num	8	YNMFMT.		Other skeletal condition
29	IHxALGSRenalOther	Num	8	YNMFMT.		Other renal structural anomaly
30	IHxAnyRenalAbnormal	Num	8	YNMFMT.		Renal abnormality
31	DrainAny	Num	8	YNMFMT.		Biliary drainage procedure
32	DrainKasai	Num	8	YNMFMT.		Kasai
33	SpleenSize	Num	8			Spleen size (cm) below costal margin (set to 0 if not palpable)
34	HeightCm	Num	8			Height (cm)
35	WeightKg	Num	8			Weight (kg)
36	Pruritus	Num	8	PRURITUSFMT.		Pruritus
37	Xanthoma	Num	8	XANTHOMAFMT.		Xanthoma

Num	Variable	Type	Len	Format	Informat	Label
38	INR	Num	8			INR
39	AlbuminGdl	Num	8			Albumin (g/dL)
40	SerumBileAcidsUmolL	Num	8			Total serum bile acids (umol/L)
41	CholesterolMgdl	Num	8			Total cholesterol (mg/dL)
42	VitaminAUgdl	Num	8			Vitamin A (ug/dL)
43	VitaminD125OH2Pgml	Num	8			1,25 OH ₂ Vitamin D (pg/mL)
44	VitaminD250HNgml	Num	8			Vitamin D (D25OH) (ng/mL)
45	VitaminEUgdl	Num	8			Vitamin E (ug/mL)
46	ALTUnitsL	Num	8			ALT (U/L)
47	ASTUnitsL	Num	8			AST (U/L)
48	GGTPUnitsL	Num	8			GGTP (U/L)
49	PlateletsCnt	Num	8			Platelets (10 ³ /mm ³)
50	BiliTotalMgdl	Num	8			Total or (unconjugated + conjugated) bilirubin (mg/dL)
51	Baseline	Num	8	YNMFMT.		Baseline
52	AgeAtExam	Num	8			Age at exam (years)
53	TimeFromSymptomOnset	Num	8			Time (years) from onset of first disease symptoms
54	Visit	Num	8	VISITFMT.		Study visit
55	AgeATxp	Num	8			Age (years) at liver transplant
56	AgeAtBaseline	Num	8			Age (years) at baseline LOGIC visit
57	JAG1	Num	8	YNMFMT.		JAG1 mutation
58	NOTCH2	Num	8	YNMFMT.		NOTCH2 mutation
59	ATP8B1	Num	8	YNMFMT.		ATP8B1 mutation
60	ABCB11	Num	8	YNMFMT.		ABCB11 mutation
61	ABCB4	Num	8	YNMFMT.		ABCB4 mutation
62	JAG1DiseaseClass1	Num	8	MUTATIONCLASSFMT.		JAG1 mutation classification
63	NOTCH2DiseaseClass1	Num	8	MUTATIONCLASSFMT.		NOTCH2 mutation classification
64	ALGSMutation	Char	35			ALGS mutation
65	One	Num	8			Variable with value of 1
66	WholeAgeAtExam	Num	8			Age at exam (years - integer portion)
67	Log10Bili	Num	8			log 10 (bilirubin)
68	Log10ALT	Num	8			log 10 (ALT)
69	Log10AST	Num	8			log 10 (AST)
70	Log10GGTP	Num	8			log 10 (GGTP)
71	Log10Cholesterol	Num	8			log 10 (Cholesterol)
72	APRI	Num	8			AST to platelet ratio index
73	SevereHeartDefect	Num	8	YNMFMT.		Severe heart defect
74	HeartDefectCat	Num	8	HEARTDEFECTCATFMT.		Heart defect category
75	ALGSMutationClass	Num	8	MUTATIONCLASSFMT.		ALGS mutation classification
76	T1	Num	8			Days from baseline

Num	Variable	Type	Len	Format	Informat	Label
77	T1Years	Num	8			Years from baseline
78	TestDone	Num	8	YNMFMT.		Audiology testing performed
79	TympDone	Num	8	TYMPFMT.		Was tympanometry done?
80	TympRE	Num	8	PPFMT.		Typanometry right ear
81	TympREFR	Num	8	FAILRSNFMT.		Right ear, reason for failure
82	TympREFRText	Char	50	\$50.		Right ear, reason for failure text
83	TympLE	Num	8	PPFMT.		Typanometry left ear
84	TympLEFR	Num	8	FAILRSNFMT.		Left ear, reason for failure
85	TympLEFRText	Char	50	\$50.		Left ear, reason for failure text
86	SurvTmTxp	Num	8			Survival time from baseline to transplant or death (days)
87	SurvTmTxpYrs	Num	8			Survival time from baseline to transplant or death (years)
88	EventTxp	Num	8			Transplant event (1= transplanted 0=censored)
89	SurvTmPat	Num	8			Survival time from baseline to death (days)
90	SurvTmPatYrs	Num	8			Survival time from baseline to death (years)
91	EventDeath	Num	8			Death event (1=died 0=censored)
92	EventTxpDeath	Num	8			Transplant or death event (1=died or transplanted 0=censored)
93	SurvAgeTxp	Num	8			Survival time from birth to transplant or death (days)
94	SurvAgeTxpYrs	Num	8			Survival time from birth to transplant or death (years)
95	T1AgeYrs	Num	8			Age at entry to risk set (years)
96	SurvAgeTxp5Yrs	Num	8			Survival time from birth to transplant or death (capped at 5 years)
97	EventTxpDeathLe5Yrs	Num	8			Transplant or death event by age 5 (1=died or transplanted 0=censored)
98	AscitesStatus	Num	8	OUTCOMESTATUSFMT.		Ascites status
99	AscitesAge	Num	8			Age (years) at ascites
100	GIBleedStatus	Num	8	OUTCOMESTATUSFMT.		GI bleed status
101	GIBleedAge	Num	8			Age (years) at GI bleed
102	PPHStatus	Num	8	OUTCOMESTATUSFMT.		PPH status
103	PPHAge	Num	8			Age (years) at PPH
104	HPSStatus	Num	8	OUTCOMESTATUSFMT.		HPS status
105	HPSAge	Num	8			Age (years) at HPS
106	SplenomegalyStatus	Num	8	OUTCOMESTATUSFMT.		Splenomegaly status
107	SplenomegalyAge	Num	8			Age (years) at splenomegaly
108	ThrombocytopeniaStatus	Num	8	OUTCOMESTATUSFMT.		Thrombocytopenia status
109	ThrombocytopeniaAge	Num	8			Age at thrombocytopenia
110	BoneFractureStatus	Num	8	OUTCOMESTATUSFMT.		Bone fracture status
111	BoneFractureAge	Num	8			Age (years) at bone fracture
112	PartBilDiversion	Char	22			Partial biliary diversion
113	IlealEx	Char	22			Ileal exclusion
114	AgeAtBaselineDisplay	Num	8			Age at baseline (years)

Num	Variable	Type	Len	Format	Informat	Label
115	HeightZScoreDisplay	Num	8			Height Z score
116	WeightZScoreDisplay	Num	8			Weight Z score
117	BMIZScoreDisplay	Num	8			BMI Z score
118	ConductionAllPass	Num	8	YNMFMT.		Passed all conduction tests
119	ConductionOneFail	Num	8	YNMFMT.		Failed at least one conduction test
120	TxpCompNone	Num	8	YNMFMT.		Complications noted at transplant - none
121	TxpCompFailThrive	Num	8	YNMFMT.		Complications noted at transplant - failure to thrive
122	TxpCompAscites	Num	8	YNMFMT.		Complications noted at transplant - ascites
123	TxpCompCholangitis	Num	8	YNMFMT.		Complications noted at transplant - cholangitis
124	TxpCompCoagulopathy	Num	8	YNMFMT.		Complications noted at transplant - coagulopathy
125	TxpCompVarices	Num	8	YNMFMT.		Complications noted at transplant - varices
126	TxpCompGIBleed	Num	8	YNMFMT.		Complications noted at transplant - GI bleed
127	TxpCompEncephalopathy	Num	8	YNMFMT.		Complications noted at transplant - encephalopathy
128	TxpCompHepatopulmonary	Num	8	YNMFMT.		Complications noted at transplant - hepatopulmonary syndrome
129	TxpCompHepatorenal	Num	8	YNMFMT.		Complications noted at transplant - hepatorenal syndrome
130	TxpCompFracture	Num	8	YNMFMT.		Complications noted at transplant - bone fracture
131	TxpCompXanthomatosis	Num	8	YNMFMT.		Complications noted at transplant - xanthomatosis
132	TxpCompPruritus	Num	8	YNMFMT.		Complications noted at transplant - pruritus
133	TxpCompOther	Num	8	YNMFMT.		Complications noted at transplant - other
134	TxpCompOSTXT	Char	200			Complications noted at transplant - other text
135	DeathCauseText	Char	200			Cause of death
136	MinBaselineDt	Num	8	MMDDYY10.		Min baseline
137	MaxBaselineDt	Num	8	MMDDYY10.		Max baseline
138	MaxFuDt	Num	8	MMDDYY10.		Max follow-up date
139	MaxFuAge	Num	8			Oldest follow up age

Data Set Name: algsformats.sas7bdat

Num	Variable	Type	Len	Label
1	FMTNAME	Char	32	Format name
2	START	Char	16	Starting value for format
3	END	Char	16	Ending value for format
4	LABEL	Char	113	Format value label
5	MIN	Num	3	Minimum length
6	MAX	Num	3	Maximum length
7	DEFAULT	Num	3	Default length
8	LENGTH	Num	3	Format length
9	FUZZ	Num	8	Fuzz value
10	PREFIX	Char	2	Prefix characters
11	MULT	Num	8	Multiplier
12	FILL	Char	1	Fill character
13	NOEDIT	Num	3	Is picture string noedit?
14	TYPE	Char	1	Type of format
15	SEXCL	Char	1	Start exclusion
16	EEXCL	Char	1	End exclusion
17	HLO	Char	13	Additional information
18	DECSEP	Char	1	Decimal separator
19	DIG3SEP	Char	1	Three-digit separator
20	DATATYPE	Char	8	Date/time/datetime?
21	LANGUAGE	Char	8	Language for date strings

Data Set Name: algsfu.sas7bdatt

Num	Variable	Type	Len	Format	Informat	Label
1	agemos	Num	8			Age at exam (months)
2	sex	Num	3	SEXFMT.		Sex
3	bmi	Num	8			BMI
4	WeightZScore	Num	8			weight-for-age Z
5	BMIZScore	Num	8			BMI-for-age Z
6	HeightZScore	Num	8			height-for-age Z
7	Ethnicity	Num	8	ETHNICITYFMT.	6.	Ethnicity
8	SubjectID	Char	8			LOGIC subject ID
9	Race	Num	8	RACEFMT.		Race
10	IHxKasai	Num	8	YNMFMT.		Kasai (initial history)
11	IHxSurgeryDrainAny	Num	8	YNMFMT.		Drainage procedure (initial history)
12	IHxALGSPPS	Num	8	YNMFMT.		Peripheral pulmonary stenosis
13	IHxALGSPulmStenosis	Num	8	YNMFMT.		Pulmonary valve stenosis
14	IHxALGSTetraFallot	Num	8	YNMFMT.		Tetralogy of Fallot
15	IHxALGSVentricDef	Num	8	YNMFMT.		Ventricular septal defect
16	IHxALGSAtriaDef	Num	8	YNMFMT.		Atrial septal defect
17	IHxALGSDepEyes	Num	8	YNMFMT.		Deep-set eyes
18	IHxALGSForehead	Num	8	YNMFMT.		Broad forehead
19	IHxALGSChin	Num	8	YNMFMT.		Pointed chin
20	IHxALGSPostEmbrotaxon	Num	8	YNMFMT.		Posterior embryotoxon
21	IHxALGSAxenfeld	Num	8	YNMFMT.		Axenfeld's anomaly
22	IHxALGSButterfly	Num	8	YNMFMT.		Butterfly vertebrae
23	IHxALGSDyspKidney	Num	8	YNMFMT.		Dysplastic kidney
24	IHxALGSSingleKidney	Num	8	YNMFMT.		Single kidney
25	IHxALGSRTA	Num	8	YNMFMT.		Renal tubular acidosis
26	IHxALGSHeartOther	Num	8	YNMFMT.		Other cardiac anomaly
27	IHxALGSEyesOther	Num	8	YNMFMT.		Other eye condition
28	IHxALGSSkelOther	Num	8	YNMFMT.		Other skeletal condition
29	IHxALGSRenalOther	Num	8	YNMFMT.		Other renal structural anomaly
30	IHxAnyRenalAbnormal	Num	8	YNMFMT.		Renal abnormality
31	DrainAny	Num	8	YNMFMT.		Biliary drainage procedure
32	DrainKasai	Num	8	YNMFMT.		Kasai
33	SpleenSize	Num	8			Spleen size (cm) below costal margin (set to 0 if not palpable)
34	HeightCm	Num	8			Height (cm)
35	WeightKg	Num	8			Weight (kg)
36	Pruritus	Num	8	PRURITUSFMT.		Pruritus
37	Xanthoma	Num	8	XANTHOMAFMT.		Xanthoma

Num	Variable	Type	Len	Format	Informat	Label
38	INR	Num	8			INR
39	AlbuminGdl	Num	8			Albumin (g/dL)
40	SerumBileAcidsUmolL	Num	8			Total serum bile acids (umol/L)
41	CholesterolMgdl	Num	8			Total cholesterol (mg/dL)
42	VitaminAUgdl	Num	8			Vitamin A (ug/dL)
43	VitaminD125OH2Pgml	Num	8			1,25 OH ₂ Vitamin D (pg/mL)
44	VitaminD250HNgml	Num	8			Vitamin D (D25OH) (ng/mL)
45	VitaminEUgdl	Num	8			Vitamin E (ug/mL)
46	ALTUnitsL	Num	8			ALT (U/L)
47	ASTUnitsL	Num	8			AST (U/L)
48	GGTPUnitsL	Num	8			GGTP (U/L)
49	PlateletsCnt	Num	8			Platelets (10 ³ /mm ³)
50	BiliTotalMgdl	Num	8			Total or (unconjugated + conjugated) bilirubin (mg/dL)
51	Baseline	Num	8	YNMFMT.		Baseline
52	AgeAtExam	Num	8			Age at exam (years)
53	TimeFromSymptomOnset	Num	8			Time (years) from onset of first disease symptoms
54	Visit	Num	8	VISITFMT.		Study visit
55	AgeAtTxp	Num	8			Age (years) at liver transplant
56	AgeAtBaseline	Num	8			Age (years) at baseline LOGIC visit
57	JAG1	Num	8	YNMFMT.		JAG1 mutation
58	NOTCH2	Num	8	YNMFMT.		NOTCH2 mutation
59	ATP8B1	Num	8	YNMFMT.		ATP8B1 mutation
60	ABCB11	Num	8	YNMFMT.		ABCB11 mutation
61	ABCB4	Num	8	YNMFMT.		ABCB4 mutation
62	JAG1DiseaseClass1	Num	8	MUTATIONCLASSFMT.		JAG1 mutation classification
63	NOTCH2DiseaseClass1	Num	8	MUTATIONCLASSFMT.		NOTCH2 mutation classification
64	ALGSMutation	Char	35			ALGS mutation
65	One	Num	8			Variable with value of 1
66	WholeAgeAtExam	Num	8			Age at exam (years - integer portion)
67	Log10Bili	Num	8			log 10 (bilirubin)
68	Log10ALT	Num	8			log 10 (ALT)
69	Log10AST	Num	8			log 10 (AST)
70	Log10GGTP	Num	8			log 10 (GGTP)
71	Log10Cholesterol	Num	8			log 10 (Cholesterol)
72	APRI	Num	8			AST to platelet ratio index
73	SevereHeartDefect	Num	8	YNMFMT.		Severe heart defect
74	HeartDefectCat	Num	8	HEARTDEFECTCATFMT.		Heart defect category
75	ALGSMutationClass	Num	8	MUTATIONCLASSFMT.		ALGS mutation classification

Num	Variable	Type	Len	Format	Informat	Label
76	T1	Num	8			Days from baseline
77	T1Years	Num	8			Years from baseline

Data Set Name: algsnhcontrolsbl.sas7bdat

Num	Variable	Type	Len	Format	Label
1	ParticipantID	Char	25	\$25.	ChiLDReN participant ID
2	SampWeight	Num	8		Sample weight (based on age distribution)
3	AgeAtBaseline	Num	8		Age at Baseline (years)
4	AgeGrp	Num	8	AGEGRPFMT.	Age group
5	Gender	Num	8	SEXFMT.	Sex
6	Race	Num	8	RACEFMT.	Race
7	Ethnicity	Num	8	ETHNICITYFMT.	Ethnicity
8	CSS	Num	8	CSSFMT.	Clinician scratch scale
9	PedsQLTotalParent	Num	8		PedsQL total score
10	BiliTotalMgdl	Num	8		Total bilirubin (mg/dl)
11	CholesterolMgdl	Num	8		Cholesterol (mg/dl)
12	SerumBileAcidsUmolL	Num	8		Serum bile acids (umol/L)
13	ASTUnitsL	Num	8		AST
14	ALTUnitsL	Num	8		ALT
15	GGTPUnitsL	Num	8		GGTP
16	AlbuminGdl	Num	8		Albumin (g/dl)
17	PlateletsCnt	Num	8		Platelet count (10 ³ /mm ³)
18	HeightZScore	Num	8		Height Z-score
19	WeightZScore	Num	8		Weight Z-score
20	SurvTmTxpDeathYrs	Num	8		Survival time from baseline to transplant or death (years)
21	SurvTmCEAscitesYrs	Num	8		Survival time from baseline to ascites (years)
22	SurvAgeTxpDeathYrs	Num	8		Survival time from birth to transplant or death (years)
23	EventDeath	Num	8		Death event (1=died 0=censored)
24	EventTxp	Num	8		Transplant event (1= transplanted 0=censored)
25	EventTxpDeath	Num	8		Transplant or death event (1=died or transplanted 0=censored)
26	EventCEAscites	Num	8		Clinically evident ascites (1=event 0=censored)
27	EventCEVaricealBleed	Num	8		Clinically evident variceal hemorrhage (1=event 0=censored)
28	EventDrain	Num	8		Partial biliary diversion or ileal exclusionn (1=event 0=censored)
29	EventComposite	Num	8		Earliest of transplant, death, ascites, bleed (1=event 0=censored)
30	CauseOfDeath	Char	4000	\$4000.	Cause of death

Data Set Name: algsnhcontrolsfu.sas7bdat

Num	Variable	Type	Len	Format	Label
1	ParticipantID	Char	25	\$25.	ChiLDReN participant ID
2	SampWeight	Num	8		Sample weight (based on age distribution)
3	AgeAtBaseline	Num	8		Age at Baseline (years)
4	AgeAtExam	Num	8		Age at Exam (years)
5	Visit	Num	8	VISITFMT.	Study visit
6	ExamTimeYrs	Num	8		Time (years) from baseline to exam
7	TransplantTimeYrs	Num	8		Time (years) from baseline to transplant
8	DeathTimeYrs	Num	8		Time (years) from baseline to death DrainPartBilDiversioTimeYrsTime (years) from baseline to partial biliary diversion
9	AgeGrp	Num	8	AGEGRPFMT.	Age group
10	Gender	Num	8	SEXFMT.	Sex
11	Race	Num	8	RACEFMT.	Race
12	Ethnicity	Num	8	ETHNICITYFMT.	Ethnicity
13	CSS	Num	8	CSSFMT.	Clinician scratch scale
14	BiliTotalMgdl	Num	8		Total bilirubin (mg/dl)
15	ALTUnitsL	Num	8		ALT
16	AlbuminGdl	Num	8		Albumin (g/dl)
17	PlateletsCnt	Num	8		Platelet count (10 ³ /mm ³)
18	HeightZScore	Num	8		Height Z-score
19	WeightZScore	Num	8		Weight Z-score
20	CSS_BL	Num	8		CSS at baseline
21	BiliTotal_BL	Num	8		Total bilirubin at baseline
22	ALT_BL	Num	8		ALT at baseline
23	Albumin_BL	Num	8		Albumin at baseline
24	Platelet_BL	Num	8		Platelet count at baseline
25	HeightZ_BL	Num	8		Height Z-score at baseline
26	WeightZ_BL	Num	8		Weight Z-score at baseline
27	CSS_Change	Num	8		CSS change from baseline
28	BiliTotal_Change	Num	8		Total bilirubin change from baseline
29	ALT_Change	Num	8		ALT change from baseline
30	Albumin_Change	Num	8		Albumin change from baseline
31	Platelet_Change	Num	8		Platelet count change from baseline
32	HeightZ_Change	Num	8		Height Z-score change from baseline
33	WeightZ_Change	Num	8		Weight Z-score change from baseline
34	CSS_ClinSig	Num	8		Clinically significant change in CSS
35	BiliTotal_ClinSig	Num	8		Clinically significant change in total bilirubin
36	ALT_ClinSig	Num	8		Clinically significant change in ALT
37	Platelet_ClinSig	Num	8		Clinically significant change in platelet count

Num	Variable	Type	Len	Format	Label
38	Albumin_ClinSig	Num	8		Clinically significant change in Alubmin
39	WeightZ_ClinSig	Num	8		Clinically significant change in weight Z-score
40	HeightZ_ClinSig	Num	8		Clinically significant change in height Z-score
41	Urso_Flag	Num	8		Ursodeoxycholic acid use noted (1=yes, 0=no)
42	Rifampin_Flag	Num	8		Rifampin use noted (1=yes, 0=no)
43	Antihist_Flag	Num	8		Antihistamine use noted (1=yes, 0=no)
44	Cholestyrya_Flag	Num	8		Cholestyramine use noted (1=yes, 0=no)
45	Phenobarb_Flag	Num	8		Phenobarbital use noted (1=yes, 0=no)
46	NonOralNutrition_Flag	Num	8		Non-oral nutrition use noted (1=yes, 0=no)
47	Naltrexone_Flag	Num	8		Naltrexone use noted (1=yes, 0=no)
48	DrainPartBilDiversionTimeYrs	Num	8		
49	DrainIlealExTimeYrs	Num	8		Time (years) from baseline to ileal exclusion

Data Set Name: algnhcontrolselect.sas7bdat

Num	Variable	Type	Len	Format	Label
1	ParticipantID	Char	25	\$25.	ChiLDReN participant ID
2	Diagnosis	Num	8	DIAGNOSISFMT.	LOGIC diagnosis
3	IncludeStudyDate	Num	8		LOGIC baseline before 1/1/2020 (1=yes, 0=no)
4	IncludePreTxp	Num	8		Pre-transplant enrollment flag (1=Pre-transplant, 0=Post-transplant)
5	IncludeDisease	Num	8		Does participant have disease? (1=yes, 0=no (unaffected sibling))
6	IncludeAge	Num	8		Age 1 to 18 years (1=yes, 0=no)
7	StudyAgeInclude	Num	8		Turned 1 year old during LOGIC study (1=yes, 0=no)
8	IncludeAnyFollowUp	Num	8		Has some follow-up information (1=yes, 0=no)
9	IncludeCholestasis	Num	8		Evidence of cholestasis (1=yes, 0=no)
10	Cholestasis1	Num	8		Serum bile acids > 50 (1=yes, 0=no)
11	Cholestasis2	Num	8		Direct bilirubin > 1 (1=yes, 0=no)
12	Cholestasis3	Num	8		Conjugated bilirubin > 1 (1=yes, 0=no)
13	Cholestasis4	Num	8		GGTP > 150 (1=yes, 0=no)
14	Cholestasis5	Num	8		CSS 3 or 4 (1=yes, 0=no)
15	IncludeScratch	Num	8		CSS 2 or higher at baseline (1=yes, 0=no)
16	ExcludeDrugTrial	Num	8		Participant in Drug Trial (1=yes, 0=no)
17	ExcludeKasai	Num	8		History of Kasai portoenterostomy (1=yes, 0=no)
18	ExcludeDrain	Num	8		History of biliary diversion or ileal exclusion (1=yes, 0=no)
19	ExcludeOtherLiverDgn	Num	8		Other liver disease noted (1=yes, 0=no)
20	ExcludeALT	Num	8		ALT over 600 (15 x ULN) (1=yes, 0=no)
21	ExcludeAlbumin	Num	8		Albumin under 3 (1=yes, 0=no)
22	ExcludeCEAscites	Num	8		History of clinically evident ascites (1=yes, 0=no)
23	ExcludeGIBleed	Num	8		History of GI bleed (1=yes, 0=no)
24	ExcludeTxpList	Num	8		History of liver transplant waiting list (1=yes, 0=no)
25	IncludeFlag	Num	8		Included in study (1=yes, 0=no)
26	MinConsent	Num	8	MMDDYY10.	Earliest eligible LOGIC consent date
27	MaxConsent	Num	8	MMDDYY10.	Most recent eligible LOGIC consent date

Data Set Name: child_univariate.sas7bdat

Num	Variable	Type	Len	Label
1	CTXSTDID	Num	8	De-Identified Subject ID
2	PHYSICAL	Num	8	QOL Physical
3	PSYCHOSOCIAL	Num	8	QOL psychosocial
4	TOTAL	Num	8	QOL Total
5	WAZ	Num	8	Weight z-scores
6	HAZ	Num	8	Height z-scores
7	BT_N	Num	8	Total Bilirubin (mg/dL)
8	DIS_GRP	Char	25	Disease Group - ALGS, BAD, PFIC, a1-AT
9	GRP	Num	8	Eligibility group

Data Set Name: demographics.sas7bdat

Num	Variable	Type	Len	Label
1	CTXSTDID	Num	8	De-Identified Subject ID
2	PHYSICAL	Num	8	QOL Physical
3	TOTAL	Num	8	QOL Total
4	WAZ	Num	8	Weight z-scores
5	HAZ	Num	8	Height z-scores
6	BT_N	Num	8	Total Bilirubin (mg/dL)
7	GGTP_N	Num	8	GGTP (U/L)
8	INR_N	Num	8	INR
9	WBC_N	Num	8	WBC (10 ³ /mm ³)
10	PLATELETS_N	Num	8	Platelet (10 ³ /mm ³)
11	ALT_N	Num	8	ALT (U/L)
12	ALBUMIN_N	Num	8	Albumin (g/dL)
13	RACE	Char	12	Race
14	GENDER	Char	6	Gender - 1=male 2=female
15	TX_LISTING	Char	1	Transplant Listing - 1=Listed for Transplant
16	DIS_GRP	Char	25	Disease Group - ALGS, BAD, PFIC, a1-AT
17	GRP	Num	8	Eligibility group
18	AGE_AT_TEST	Num	8	Age at QOL survey (years)

Data Set Name: dxadata.sas7bdat

Num	Variable	Type	Len	Label
1	BMDSPINE	Num	8	Total spine BMD
2	BMDSPINEZSCORE	Num	8	Total spine BMD Z-score
3	BMDNECK	Num	8	Hip neck BMD
4	BMDNECKZSCORE	Num	8	Hip neck BMD Z-score
5	BMDHIP	Num	8	Total hip BMD
6	BMDHIPZSCORE	Num	8	Total hip BMD Z-score
7	BMDSPINEWTHTFATZSCORE	Num	8	Total spine BMD Z-score adjusted for weight, height, fat
8	BMDNECKWTHTFATZSCORE	Num	8	Hip neck BMD Z-score adjusted for weight, height, fat
9	BMDHIPWTHTFATZSCORE	Num	8	Total hip BMD Z-score adjusted for weight, height, fat
10	BMCTOTALWTHTFATZSCORE	Num	8	Whole body BMC Z-score adjusted for weight, height, fat
11	BMDTOTALWTHTFATZSCORE	Num	8	Whole body BMD Z-score adjusted for weight, height, fat
12	BMCSPINEWTHTFATZSCORE	Num	8	Total spine BMC Z-score adjusted for weight, height, fat
13	BMDSPINEWHTZSCORE	Num	8	Total spine BMD Z-score adjusted for weight, height
14	BMDNECKWHTZSCORE	Num	8	Hip neck BMD Z-score adjusted for weight, height
15	BMDHIPWHTZSCORE	Num	8	Total hip BMD Z-score adjusted for weight, height
16	BMCTOTALWHTZSCORE	Num	8	Whole body BMC Z-score adjusted for weight, height
17	BMDTOTALWHTZSCORE	Num	8	Whole body BMD Z-score adjusted for weight, height
18	BMCSPINEWHTZSCORE	Num	8	Total spine BMC Z-score adjusted for weight, height
19	MEASUREDPCTLEAN	Num	8	Measured lean body mass (%)
20	BMCTOTAL	Num	8	Whole body BMC
21	BMDTOTAL	Num	8	Whole body BMD
22	BMDSUBTOTAL	Num	8	Whole body minus head BMD
23	BMCSUBTOTAL	Num	8	Whole body minus head BMC
24	BMDTOTALZSCORE	Num	8	Whole body BMD Z-score
25	BMDSUBTOTALZSCORE	Num	8	Whole body minus head BMD Z-score
26	BMCTOTALZSCORE	Num	8	Whole body BMC Z-score
27	BMCSUBTOTALZSCORE	Num	8	Whole body minus head BMC Z-score
28	BMCSUBTOTALWTHTFATZSCORE	Num	8	Whole body minus head BMC Z-score adjusted for weight, height, fat
29	BMDSUBTOTALWTHTFATZSCORE	Num	8	Whole body minus head BMD Z-score adjusted for weight, height, fat
30	BMCNECKWTHTFATZSCORE	Num	8	Hip neck BMC Z-score adjusted for weight, height, fat
31	BMCHIPWTHTFATZSCORE	Num	8	Total hip BMC Z-score adjusted for weight, height, fat
32	BMCSUBTOTALWHTZSCORE	Num	8	Whole body minus head BMC Z-score adjusted for weight, height
33	BMDSUBTOTALWHTZSCORE	Num	8	Whole body minus head BMD Z-score adjusted for weight, height
34	BMCNECKWHTZSCORE	Num	8	Hip neck BMC Z-score adjusted for weight, height
35	BMCHIPWHTZSCORE	Num	8	Total hip BMC Z-score adjusted for weight, height
36	SUBJECTID	Char	7	LOGIC subject ID
37	LOGICGROUP	Num	8	LOGIC study group

Num	Variable	Type	Len	Label
38	NODXA	Num	8	Flag for subjects without a scan used as matched comparison (0=DXA scan, 1=no DXA scan)
39	DGNDETAIL	Num	8	Detailed diagnosis from diagnosis CRF
40	DIAGNOSIS	Num	8	Diagnosis group
41	SEX	Num	8	Gender
42	ETHNICITY	Num	8	Ethnicity
43	RACE	Num	8	Race
44	AGEATSCANYEARS	Num	8	Age (years) at the time of scan
45	AGEATBASELINEYEARS	Num	8	Age (years) at LOGIC study baseline
46	AGEATSCANORMATCH	Num	8	Age (years) at DXA scan or date used to match non-DXA subject
47	HEIGHTCM	Num	8	Height (cm) closest to scan
48	WEIGHTKG	Num	8	Weight (kg) closest to scan
49	SPLEENSIZE	Num	8	Spleen size (cm below costal margin)
50	BILITOTAL	Num	8	Total bilirubin (mg/dL)
51	BILIDIRECT	Num	8	Direct bilirubin (mg/dL)
52	ALBUMIN	Num	8	Albumin (g/dL)
53	BILEACIDUM	Num	8	Total serum bile acids (umol/L)
54	BILEACIDUG	Num	8	Total serum bile acids (ug/dL)
55	INR	Num	8	INR
56	VITAMIND	Num	8	Vitamin D (ng/mL)
57	ALKPHOS	Num	8	Alkaline phosphatase (U/L)
58	GGT	Num	8	GGT (U/L)
59	ALT	Num	8	ALT (U/L)
60	AST	Num	8	AST (U/L)
61	WBC	Num	8	WBC
62	HGB	Num	8	Hemoglobin (g/dl)
63	PLATE	Num	8	Platelets
64	BONEFRACTUREANY	Num	8	any bone fracture
65	BONEFRACTUREGT1	Num	8	more than one bone fracture
66	BONEFRACTURENUM	Num	8	Number of bone fractures
67	DIVERSIONFLAG	Num	8	Biliary diversion prior to scan
68	PHYSICALEXAMAGEYEARS	Num	8	Age (years) at the time of height/weight measurement
69	PHYSICALEXAMAGEMONTHS	Num	8	Age (months) at the time of height/weight measurement
70	BMI	Num	8	BMI closest to scan
71	WEIGHTZSCORE	Num	8	Weight Z-score
72	BMIZSCORE	Num	8	BMI Z-score
73	HEIGHTZSCORE	Num	8	Height Z-Score
74	SCANYEAR	Num	8	Year of DXA scan
75	LEANBODYMASSKG	Num	8	Calculated lean body mass (kg)

Num	Variable	Type	Len	Label
76	LEANBODYMASSPERCENT	Num	8	Calculated lean body mass (percent)
77	DIVERSIONTOSCANYRS	Num	8	Time (years) from biliary diversion to DXA scan
78	DIAGNOSISORDER	Num	8	Alternate ordering of diagnosis categories
79	BMDTOTALZSCORE_ORIG	Num	8	Non-Winsorized Whole body BMD Z-score
80	BMDTOTALWHTZSCORE_ORIG	Num	8	Non-Winsorized Whole body BMD Z-score adjusted for weight, height
81	BMDTOTALWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Whole body BMD Z-score adjusted for weight, height, fat
82	BMCTOTALZSCORE_ORIG	Num	8	Non-Winsorized Whole body BMC Z-score
83	BMCTOTALWHTZSCORE_ORIG	Num	8	Non-Winsorized Whole body BMC Z-score adjusted for weight, height
84	BMCTOTALWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Whole body BMC Z-score adjusted for weight, height, fat
85	BMDSUBTOTALZSCORE_ORIG	Num	8	Non-Winsorized Whole body minus head BMD Z-score
86	BMDSUBTOTALWHTZSCORE_ORIG	Num	8	Non-Winsorized Whole body minus head BMD Z-score adjusted for weight, height
87	BMDSUBTOTALWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Whole body minus head BMD Z-score adjusted for weight, height, fat
88	BMCSUBTOTALZSCORE_ORIG	Num	8	Non-Winsorized Whole body minus head BMC Z-score
89	BMCSUBTOTALWHTZSCORE_ORIG	Num	8	Non-Winsorized Whole body minus head BMC Z-score adjusted for weight, height
90	BMCSUBTOTALWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Whole body minus head BMC Z-score adjusted for weight, height, fat
91	BMDSPINEZSCORE_ORIG	Num	8	Non-Winsorized Total spine BMD Z-score
92	BMDSPINEWHTZSCORE_ORIG	Num	8	Non-Winsorized Total spine BMD Z-score adjusted for weight, height
93	BMDSPINEWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Total spine BMD Z-score adjusted for weight, height, fat
94	BMCSPINEWHTZSCORE_ORIG	Num	8	Non-Winsorized Total spine BMC Z-score adjusted for weight, height
95	BMCSPINEWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Total spine BMC Z-score adjusted for weight, height, fat
96	BMDNECKZSCORE_ORIG	Num	8	Non-Winsorized Hip neck BMD Z-score
97	BMDNECKWHTZSCORE_ORIG	Num	8	Non-Winsorized Hip neck BMD Z-score adjusted for weight, height
98	BMDNECKWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Hip neck BMD Z-score adjusted for weight, height, fat
99	BMCNECKWHTZSCORE_ORIG	Num	8	Non-Winsorized Hip neck BMC Z-score adjusted for weight, height
100	BMCNECKWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Hip neck BMC Z-score adjusted for weight, height, fat
101	BMDHIPZSCORE_ORIG	Num	8	Non-Winsorized Total hip BMD Z-score
102	BMDHIPWHTZSCORE_ORIG	Num	8	Non-Winsorized Total hip BMD Z-score adjusted for weight, height
103	BMDHIPWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Total hip BMD Z-score adjusted for weight, height, fat
104	BMCHIPWHTZSCORE_ORIG	Num	8	Non-Winsorized Total hip BMC Z-score adjusted for weight, height
105	BMCHIPWHTFATZSCORE_ORIG	Num	8	Non-Winsorized Total hip BMC Z-score adjusted for weight, height, fat
106	BMDTOTALZSCORE_WINFLAG	Num	8	Was BMDTotalZScore Winsorized?
107	BMDTOTALWHTZSCORE_WINFLAG	Num	8	Was BMDTotalWtHtZScore Winsorized?
108	BMDTOTALWHTFATZSCORE_WINFLAG	Num	8	Was BMDTotalWtHtFatZScore Winsorized?
109	BMCTOTALZSCORE_WINFLAG	Num	8	Was BMCTotalZScore Winsorized?
110	BMCTOTALWHTZSCORE_WINFLAG	Num	8	Was BMCTotalWtHtZScore Winsorized?
111	BMCTOTALWHTFATZSCORE_WINFLAG	Num	8	Was BMCTotalWtHtFatZScore Winsorized?
112	BMDSUBTOTALZSCORE_WINFLAG	Num	8	Was BMDSubTotalZScore Winsorized?
113	BMDSUBTOTALWHTZSCORE_WINFLAG	Num	8	Was BMDSubTotalWtHtZScore Winsorized?
114	BMDSUBTOTALWHTFATZSCORE_WINFLAG	Num	8	Was BMDSubTotalWtHtFatZScore Winsorized?

Num	Variable	Type	Len	Label
115	BMCSUBTOTALZSCORE_WINFLAG	Num	8	Was BMCSubTotalZScore Winsorized?
116	BMCSUBTOTALWTHTZSCORE_WINFLAG	Num	8	Was BMCSubTotalWtHtZScore Winsorized?
117	BMCSUBTOTALWTHTFATZSCORE_WINFLAG	Num	8	Was BMCSubTotalWtHtFatZScore Winsorized?
118	BMDSPINEZSCORE_WINFLAG	Num	8	Was BMDSpineZScore Winsorized?
119	BMDSPINEWTHTZSCORE_WINFLAG	Num	8	Was BMDSpineWtHtZScore Winsorized?
120	BMDSPINEWTHTFATZSCORE_WINFLAG	Num	8	Was BMDSpineWtHtFatZScore Winsorized?
121	BMCSPINEWTHTZSCORE_WINFLAG	Num	8	Was BMCSpineWtHtZScore Winsorized?
122	BMCSPINEWTHTFATZSCORE_WINFLAG	Num	8	Was BMCSpineWtHtFatZScore Winsorized?
123	BMDNECKZSCORE_WINFLAG	Num	8	Was BMDNeckZScore Winsorized?
124	BMDNECKWTHTZSCORE_WINFLAG	Num	8	Was BMDNeckWtHtZScore Winsorized?
125	BMDNECKWTHTFATZSCORE_WINFLAG	Num	8	Was BMDNeckWtHtFatZScore Winsorized?
126	BMCNECKWTHTZSCORE_WINFLAG	Num	8	Was BMCNeckWtHtZScore Winsorized?
127	BMCNECKWTHTFATZSCORE_WINFLAG	Num	8	Was BMCNeckWtHtFatZScore Winsorized?
128	BMDHIPZSCORE_WINFLAG	Num	8	Was BMDHipZScore Winsorized?
129	BMDHIPWTHTZSCORE_WINFLAG	Num	8	Was BMDHipWtHtZScore Winsorized?
130	BMDHIPWTHTFATZSCORE_WINFLAG	Num	8	Was BMDHipWtHtFatZScore Winsorized?
131	BMCHIPWTHTZSCORE_WINFLAG	Num	8	Was BMCHipWtHtZScore Winsorized?
132	BMCHIPWTHTFATZSCORE_WINFLAG	Num	8	Was BMCHipWtHtFatZScore Winsorized?
133	TANNERSTAGEDEVELOP	Num	8	Tanner score - development
134	TANNERSTAGEHAIR	Num	8	Tanner score - pubic hair
135	SPLEENPLATE	Num	8	Spleen > 2cm and Platelet count <150
136	BMILTNEG2	Num	8	BMI Z-score < -2
137	BONERISKYEARS	Num	8	Time (years) at risk for bone fractures
138	CENTRALBILEACID	Num	8	Total serum bile acids (umol/L)
139	GENETICDGN	Num	8	Genetic diagnosis

Data Set Name: manuscript_data_03oct19.sas7bdat

Num	Variable	Type	Len	Format	Informat	Label
1	ChiLDReN_StudyID	Char	50	\$50.	\$50.	ChiLDReN_StudyID
2	sex	Num	8	SEXF.	6.	Sex
3	Ethnicity	Num	8	ETHNF.	6.	Ethnicity
4	dx	Num	8	DXF.		Diagnosis
5	age_consent_y	Num	8			Age at consent (years)
6	VCI	Num	8	11.		Verbal Comprehension
7	PRI	Num	8	11.		Perceptual Reasoning
8	WMI	Num	8	11.		Working Memory
9	PSI	Num	8	11.		Processing Speed
10	FSIQ	Num	8	11.		Full Scale IQ
11	VIQ	Num	8	11.		Verbal IQ
12	PIQ	Num	8	11.		Performance IQ
13	GLC	Num	8	11.		General Language Component
14	age_test_y	Num	8			Age at testing (years)
15	race	Num	8	RACEF.		Race
16	CEPH	Num	8	CEPHF.		CEPH
17	college	Num	8	COLLEGEF.		Parental education
18	xanthoma	Num	8	PRESENTF.		History of xanthoma(s)
19	waz	Num	8			Weight-for-age z-score
20	bmiz	Num	8			BMI z-score
21	haz	Num	8			Height-for-age z-score
22	pruritus	Num	8	PRURITUSF.		Pruritus
23	INR	Num	8	8.1		INR
24	AlbuminGdl	Num	8			Albumin (g/dl)
25	BUNMgdl	Num	8			BUN (mg/dl)
26	CreatinineMgdl	Num	8			Creatinine (mg/dl)
27	HgbGdl	Num	8			Hemoglobin (g/dl)
28	TotalBilirubinMgdl	Num	8			Total bilirubin (mg/dl)
29	AlkPhosphUnitsL	Num	8	BEST12.		Alkaline phosphatase (U/L)
30	ALTUnitsL	Num	8	BEST12.		ALT (U/L)
31	ASTUnitsL	Num	8	BEST12.		AST (U/L)
32	GGTPUnitsL	Num	8	BEST12.		GGTP (U/L)
33	PlateletsCnt	Num	8	BEST12.		Platelet count
34	APRI	Num	8			APRI
35	FIB_4	Num	8			FIB-4
36	cardiac_cat	Num	8	CARDIAC_CATF.		Cardiac disease severity
37	JAG1	Num	8	YN.		JAG1 mutation

Num	Variable	Type	Len	Format	Informat	Label
38	jag1combo	Num	8	GENEF.		JAG1 mutation
39	ATP8B1combo	Num	8	GENEF.		ATP8B1 mutation
40	ABCB11combo	Num	8	GENEF.		ABCB11 mutation
41	ABCB4combo	Num	8	GENEF.		ABCB4 mutation
42	A1ATPhenoCombo	Num	8	PHENOF.		A1AT Phenotype
43	IHxAnyRenalAbnormal	Num	8	PRESENTF.		Any renal abnormality
44	time_fu_y	Num	8			Follow-up time (years)
45	tested	Num	8	YN.		Tested
46	test	Num	8	TESTF.		Test type

Data Set Name: parent_univariate.sas7bdat

Num	Variable	Type	Len	Label
1	CTXSTDID	Num	8	De-Identified Subject ID
2	PHYSICAL	Num	8	QOL Physical
3	PSYCHOSOCIAL	Num	8	QOL psychosocial
4	TOTAL	Num	8	QOL Total
5	WAZ	Num	8	Weight z-scores
6	HAZ	Num	8	Height z-scores
7	INR_N	Num	8	INR
8	DIS_GRP	Char	25	Disease Group - ALGS, BAD, PFIC, a1-AT
9	GRP	Num	8	Eligibility group
10	AGE_AT_TEST	Num	8	Age at QOL survey (years)
11	CARDIAC_DEFECT	Char	1	Cardiac Defect - 1=No Defect, 2=Other Defect, 3=PPS

Data Set Name: pedsq1_child_dis.sas7bdat

Num	Variable	Type	Len	Label
1	DIS_GRP	Char	25	Disease Group - ALGS, BAD, PFIC, a1-AT
2	CTXSTDID	Num	8	De-Identified Subject ID
3	GRP	Num	8	Eligibility group
4	FROM	Char	20	QOL form source
5	PHYSICAL	Num	8	QOL Physical
6	EMOTION	Num	8	QOL emotion
7	SOCIAL	Num	8	QOL social
8	SCHOOL	Num	8	QOL school
9	PSYCHOSOCIAL	Num	8	QOL psychosocial
10	TOTAL	Num	8	QOL Total

Data Set Name: pedsq_parent_dis.sas7bdat

Num	Variable	Type	Len	Label
1	DIS_GRP	Char	25	Disease Group - ALGS, BAD, PFIC, a1-AT
2	CTXSTDID	Num	8	De-Identified Subject ID
3	GRP	Num	8	Eligibility group
4	FROM	Char	21	QOL form source
5	PHYSICAL	Num	8	QOL Physical
6	EMOTION	Num	8	QOL emotion
7	SOCIAL	Num	8	QOL social
8	SCHOOL	Num	8	QOL school
9	PSYCHOSOCIAL	Num	8	QOL psychosocial
10	TOTAL	Num	8	QOL Total

Data Set Name: pfic.sas7bdat

Num	Variable	Type	Len	Format	Informat	Label
1	bmi	Num	8			BMI at baseline exam
2	WeightZScore	Num	8			weight-for-age Z at baseline exam
3	BMIZScore	Num	8			BMI-for-age Z at baeline exam
4	HeightZScore	Num	8			height-for-age Z at baeline exam
5	Gender	Num	8	SEXFMT.	6.	Sex
6	Ethnicity	Num	8	ETHNICITYFMT.	6.	Ethnicity
7	SubjectID	Char	8			LOGIC subject ID
8	PrimaryDgn	Num	8	PRIMARYDGNFMT.		Primary diagnosis
9	LogicGroup	Num	8	LOGICGROUPEFMT.		LOGIC group
10	Race	Num	8	RACEFMT.		Race
11	AgeAtConsent	Num	8			Age at consent (years)
12	DrainAny	Num	8	YESNOMISSFMT.		Any drainage procedure performed
13	DrainSelect	Num	8	YESNOMISSFMT.		Subject had Choledochojejunostomy, Biliary diversion or Ileal exclusion any time during study
14	SpleenSize	Num	8			Spleen size (cm) below costal margin (set to 0 if not palpable)
15	HeightCm	Num	8			Height (cm) at baseline exam
16	WeightKg	Num	8			Weight (kg) at baseline exam
17	Pruritus	Num	8	PRURITUSFMT.		Pruritus at baseline physical exam
18	Xanthoma	Num	8	XANTHOMAFMT.		Xanthoma at baseline exam
19	INR	Num	8			INR at baseline exam
20	AlbuminGdl	Num	8			Albumin (g/dL) at baseline exam
21	SerumBileAcidsUmolL	Num	8			Total serum bile acids (umol/L) at baseline exam
22	CalciumMgdl	Num	8			Calcium (mg/dL) at baseline exam
23	CholesterolMgdl	Num	8			Total cholesterol (mg/dL) at baseline exam
24	CreatinineMgdl	Num	8			Creatinine (mg/dL) at baseline exam
25	GlucoseMgdl	Num	8			Glucose (mg/dL) at baseline exam
26	HgbGdl	Num	8			Hemoglobin (g/dL) at baseline exam
27	PhosphorusMgdl	Num	8			Phosphorus (mg/dL) at baseline exam
28	RBPMgdl	Num	8			Retinol binding protein (mg/dL) at baseline exam
29	SerumLipidsMgdl	Num	8			Total serum lipids (mg/dL) at baseline exam
30	TotalProteinGdl	Num	8			Total protein (g/dL) at baseline exam
31	VitaminAUgdl	Num	8			Vitamin A (ug/dL) at baseline exam
32	VitaminD125OH2Pgml	Num	8			1,25 OH ₂ Vitamin D (pg/mL) at baseline exam
33	VitaminD250HNgml	Num	8			Vitamin D (D25OH) (ng/mL) at baseline exam
34	VitaminEUgdl	Num	8			Vitamin E (ug/mL) at baseline exam
35	AlkPhosphUnitsL	Num	8			Alkaline phosphate (U/L) at baseline exam
36	ALTUnitsL	Num	8			ALT (U/L) at baseline exam
37	ASTUnitsL	Num	8			AST (U/L) at baseline exam

Num	Variable	Type	Len	Format	Informat	Label
38	ClMmolL	Num	8			Choloride (mmol/L) at baseline exam
39	CO2MmolL	Num	8			Bicarbonate (mmol/L) at baseline exam
40	GGTPUnitsL	Num	8			GGTP (U/L) at baseline exam
41	KMmolL	Num	8			Potassium (mmol/L) at baseline exam
42	NaMmolL	Num	8			Sodium (mmol/L) at baseline exam
43	PlateletsCnt	Num	8			Platelets (10 ³ /mm ³) at baseline exam
44	WBCcnt	Num	8			White blood cells (10 ³ /mm ³) at baseline exam
45	BiliTotalMgdl	Num	8			Total bilirubin (mg/dL) at baseline exam
46	RBC	Num	8			Red blood cells (10 ⁶ /mm ³) at baseline exam
47	AgeAtExam	Num	8			Age at exam (years) at baseline exam
48	TimeFromSymptomOnset	Num	8			Time (years) from onset of first disease symptoms to baseline exam
49	TDDrainSelect	Num	8	YESNOMISSFMT.		Subject had Choledochojejunostomy, Biliary diversion or Ileal exclusion on or before exam
50	AgeATxp	Num	8			Age (years) at liver transplant
51	AgeAtBaseline	Num	8			Age (years) at baseline LOGIC visit
52	AgeAtFirstSymptomYrs	Num	8			Age (years) at onset of first liver disease symptoms
53	AgeAtFirstDgnYrs	Num	8			Age (years) at initial diagnosis of liver disease (from initial history CRF)
54	TxpTimeYrs	Num	8			Time (years) from LOGIC consent to liver transplant (groups 1 and 2 only)
55	GeneticDgn	Num	8	GENETICDGNFMT.		Genetic diagnosis
56	nMutations	Num	8			nMutations
57	MutationCode	Char	7	\$7.	\$7.	MutationCode
58	GeneticGroup	Num	8	GENETICGROUPEFMT.		GeneticGroup
59	dgnSpecifyOther	Char	200	\$200.		Other, specify diagnosis field
60	DaysToSelectDrain	Num	8			Number of days from baseline to drainage procedure
61	DaysToTransplant	Num	8			Number of days from baseline to transplant
62	AgeAtConsentMo	Num	8			Age (months) at enrollment
63	AgeAtFirstDgnMo	Num	8			Age (months) at diagnosis
64	AgeAtFirstSymptomMo	Num	8			Age (months) at onset of first liver disease symptoms
65	BLUrso	Num	8	YESNOMISSFMT.		Urso at baseline
66	PruritusAtBL	Num	8	YESNOMISSFMT.		History of pruritus on or before baseline exam
67	JaundiceAtBL	Num	8	YESNOMISSFMT.		History of jaundice on or before baseline exam
68	DiarrheaAtBL	Num	8	YESNOMISSFMT.		History of diarrhea on or before baseline exam
69	Failure2ThriveAtBL	Num	8	YESNOMISSFMT.		History of failure to thrive on or before baseline exam
70	BleedingAtBL	Num	8	YESNOMISSFMT.		History of bleeding or bruising on or before baseline exam
71	DiureticsAtBL	Num	8	YESNOMISSFMT.		Ascites at baseline based on diuretic use
72	GIBleedAtBL	Num	8	YESNOMISSFMT.		History of GI bleeding on or before baseline exam
73	BoneFractureAtBL	Num	8	YESNOMISSFMT.		History of bone fracture on or before baseline exam
74	RicketsAtBL	Num	8	YESNOMISSFMT.		History of rickets on or before baseline exam

Num	Variable	Type	Len	Format	Informat	Label
75	GallstonesAtBL	Num	8	YESNOMISSFMT.		History of gallstones on or before baseline exam
76	HearingProbAtBL	Num	8	YESNOMISSFMT.		History of hearing problems on or before baseline exam
77	PreBLAndTxpSelectDrain	Num	8	YESNOMISSFMT.		Drainage procedure performed prior to baseline (or transplant for post-transplant participants)
78	PruritusRxAtBL	Num	8	YESNOMISSFMT.		Pruritus Rx at baseline
79	PreBLGt1YrSelectDrain	Num	8	YESNOMISSFMT.		Drain placed at least one year before baseline
80	PerfectDrain	Num	8	YESNOMISSFMT.		Bili le 1, no pruritus meds, minimal or no pruritus, platelets at least 150, spleen lt 2cm
81	SuccessfulDrain	Num	8	YESNOMISSFMT.		Bili le 1, no pruritus meds, minimal or no pruritus, platelets at least 150, spleen lt 2cm
82	One	Num	8			Variable with value of 1
83	MutationCodeNum	Num	8	MUTATIONCODENUMFMT.		Detailed mutation group
84	FirstBaseline	Num	8	MMDDYY10.		Earliest enrollment date for participants in the study
85	LastBaseline	Num	8	MMDDYY10.		Latest enrollment date for participants in the study

Data Set Name: pficgenetics.sas7bdat

Num	Variable	Type	Len	Format	Label
1	SubjectID	Char	8		LOGIC subject ID
2	GeneticGroup	Num	8	GENETICGROUPEFMT.	GeneticGroup
3	LogicGroup	Num	8	LOGICGROUPEFMT.	LOGIC group
4	Gene	Num	8	GENEFMT.	Gene
5	MutationYN	Num	8	YNUFMT.	Mutation status
6	TestDate	Num	8	MMDDYY8.	B:1 DNA Test Date:
7	cDNAChange	Num	8	YESNOMISSFMT.	B-5: Mutation Detected : 1: cDNA change, specify:
8	Mutation1_Other	Char	200	\$50.	B-5: Mutation Detected : 1: cDNA change, specify: [text]
9	ProteinChange	Num	8	YESNOMISSFMT.	B-5: Mutation Detected : 2: Protein change, specify:
10	Mutation2_Other	Char	200	\$50.	B-5: Mutation Detected : 2: Protein change, specify: [text]
11	MutChangeType	Num	8	MUTCHANGETYPEFMT.	B:6 Change Type:
12	DiseaseCause	Num	8	DISEASECAUSEFMT.	B:7 Disease Causing:

Data Set Name: sarcopenia.sas7bdat

Num	Variable	Type	Len	Format	Label
1	ParticipantID	Char	25	\$25.	ChiLDReN participant ID
2	AgeAtScanYears	Num	8		Age (years) at the time of scan
3	PhysicalExamAgeYears	Num	8		Age (years) at the time of height/weight measurement
4	CentralBileAcid	Num	8		Bile acids (centrally tested) with outlier removed
5	LeanBodyMassKg	Num	8		Calculated lean body mass (kg)
6	LeanBodyMassPercent	Num	8		Calculated lean body mass (percent)
7	BMCTotal	Num	8		Whole body BMC
8	BMDTotal	Num	8		Whole body BMD
9	BMCTotalZScore	Num	8		Whole body BMC Z-score
10	BMDTotalZScore	Num	8		Whole body BMD Z-score
11	BMDTotalZScore_Orig	Num	8		Non-Winsorized Whole body BMD Z-score
12	BMCTotalZScore_Orig	Num	8		Non-Winsorized Whole body BMC Z-score
13	TannerStage	Num	8		Tanner stage
14	TannerStage5	Num	8		Tanner stage 5 (or 15+ years old if missing) (1=yes 0=no)
15	Sex	Num	8	SEXFMT.	Sex
16	Race	Num	8	RACEFMT.	Race
17	Ethnicity	Num	8	ETHNICITYFMT.	Ethnicity
18	Diagnosis	Num	8	DIAGNOSISFMT.	LOGIC diagnosis
19	DiversionPreDexa	Num	8		Biliary diversion prior to DEXA (1=yes 0=no)
20	GeneticDgnPFIC	Num	8	GENETICDGNPFICFMT.	PFIC genetic diagnosis
21	BoneFracturePreDexa	Num	8		Bone fracture prior to DEXA (1=yes 0=no)
22	HeightCm	Num	8		Height (cm)
23	WeightKg	Num	8		Weight (kg)
24	HeightZ	Num	8		Height Z-score
25	WeightZ	Num	8		Weight Z-score
26	BMIZ	Num	8		BMI Z-score
27	BMI	Num	8		BMI
28	BiliTotalMgdl	Num	8		Total bilirubin (mg/dL)
29	BiliDirectMgdl	Num	8		Direct bilirubin (mg/dL)
30	PedsQLTotalParent	Num	8		PedsQL total (parent)
31	PedsQLPhysParent	Num	8		PedsQL physical component (parent)
32	PedsQLTotalParticipant	Num	8		PedsQL total (participant)
33	PedsQLPhysParticipant	Num	8		PedsQL physical component (participant)
34	SpleenSizeCm	Num	8		Spleen size (cm) below costal margin
35	Albumingdl	Num	8		Albumin (g/dL)
36	INR	Num	8		INR
37	GGTPUnitsL	Num	8		GGTP

Num	Variable	Type	Len	Format	Label
38	ALTUnitsL	Num	8		ALT
39	ASTUnitsL	Num	8		AST
40	PlateletsCnt	Num	8		Platelet count ($10^3/\text{mm}^3$)
41	ALM	Num	8		Appendicular lean mass (kg)
42	FatPctTot	Num	8		Total fat %
43	FatTot	Num	8		Total Fat (gm)
44	FatTotKg	Num	8		Total Fat (kg)
45	FatIndex	Num	8		Fat mass (kg)/height (m) ²
46	ALMHeight	Num	8		ALM/Height (kg/m ²)
47	eSMM	Num	8		estimated skeletal muscle mass (kg)
48	SMPredicted	Num	8		predicted SMM
49	eSMMZ	Num	8		eSMM Z-score
50	Sarcopenia	Num	8		eSMM Z-score ≤ -2
51	eSMMHeight	Num	8		eSMM/Height (kg/m ²)
52	APRI	Num	8		AST to platelet ratio index (APRI)
53	CentralBileAcidOrig	Num	8		Bile acids (centrally tested)
54	SurvYrsTxpDeath	Num	8		Survival time (years) from DEXA for transplant/death
55	RiskTxpDeath	Num	8		In risk set for transplant/death (1=yes 0=no)
56	EventTxp	Num	8		Event indicator for transplant (1=event 0=censored)
57	EventDeath	Num	8		Event indicator for death (1=event 0=censored)
58	EventTxpDeath	Num	8		Event indicator for transplant/death (1=event 0=censored)
59	CEPHPreScan	Num	8		CEPH possible or definite prior to DEXA
60	SurvYrsComposite	Num	8		Survival time (years) from DEXA for composite outcome
61	RiskComposite	Num	8		In risk set for composite outcome (1=yes 0=no)
62	EventComposite	Num	8		Event indicator for composite outcome (1=event 0=censored)
63	BoneFractureAny	Num	8		Bone fracture before or after DEXA
64	EventCEPH	Num	8		CEPH after DEXA (1=event 0=censored)
65	EventCEVaricealBleed	Num	8		CE variceal bleed after DEXA (1=event 0=censored)
66	EventCEAscites	Num	8		CE Ascites after DEXA (1=event 0=censored)
67	EventBoneFracture	Num	8		Bone fracture after DEXA (1=event 0=censored)