

## Data Set Name: a1atbaseline.sas7bdat

Variable Number 1 Variable GROUP Type Numeric Length 8 Format . Label Eligibility Group

Variable Number 2 Variable RACECAT Type Character Length 1 Format . Label Race category

Variable Number 3 Variable DMGAB02 Type Character Length 3 Format . Label Gender

Variable Number 4 Variable DMGAB03 Type Character Length 3 Format . Label Ethnicity

Variable Number 5 Variable AST Type Numeric Length 8 Format . Label AST U/L

Variable Number 6 Variable ALT Type Numeric Length 8 Format . Label ALT U/L

Variable Number 7 Variable INR Type Numeric Length 8 Format . Label INR

Variable Number 8 Variable GGTP Type Numeric Length 8 Format . Label GGTP U/L

Variable Number 9 Variable WBC Type Numeric Length 8 Format . Label WBC 10/mm

Variable Number 10 Variable AST\_ALT\_RATIO Type Numeric Length 8 Format . Label AST/ALT Ratio

Variable Number 11 Variable PHT Type Numeric Length 8 Format . Label Portal Hypertension

Variable Number 12 Variable PHT\_US Type Numeric Length 8 Format . Label Portal Hypertension ultrasound finding

Variable Number 13 Variable LIVER\_DISEASE Type Numeric Length 8 Format . Label Liver disease severity: Mild vs. Severe

Variable Number 14 Variable AGE Type Numeric Length 8 Format . Label Age (at consent)

Variable Number 15 Variable IHX2B01\_AGE Type Numeric Length 8 Format . Label Age first noticed symptoms of liver disease

Variable Number 16 Variable IHX2B02\_AGE Type Numeric Length 8 Format . Label Age of diagnosis of CLiC liver disease

Variable Number 17 Variable ZZ Type Numeric Length 8 Format . Label ZZ Genotype/phenotype

Variable Number 18 Variable SZ Type Numeric Length 8 Format . Label SZ Genotype/phenotype

Variable Number 19 Variable PRETX Type Numeric Length 8 Format . Label Liver transplant

Variable Number 20 Variable ID Type Numeric Length 8 Format . Label De-Identified Subject ID

## Data Set Name: a1atlongv4.sas7bdat

Variable Number 1 Variable bmi Type Numeric Length 8 Format NONE Label BMI

Variable Number 2 Variable WeightZScore Type Numeric Length 8 Format NONE Label Weight-for-age Z

Variable Number 3 Variable headcz Type Numeric Length 8 Format NONE Label Head circ-for-age Z

Variable Number 4 Variable BMIZScore Type Numeric Length 8 Format NONE Label BMI-for-age Z

Variable Number 5 Variable HeightZScore Type Numeric Length 8 Format NONE Label Height-for-age Z

Variable Number 6 Variable PiZZPheno Type Numeric Length 8 Format YN. Label PiZZ phenotype

Variable Number 7 Variable PiSZPheno Type Numeric Length 8 Format YN. Label PiSZ phenotype

Variable Number 8 Variable Gender Type Numeric Length 8 Format SEX. Label Sex

Variable Number 9 Variable Ethnicity Type Numeric Length 8 Format ETHF. Label Ethnicity

Variable Number 10 Variable FinalStatus Type Numeric Length 8 Format ALLLEAVE. Label Final status

Variable Number 11 Variable SubjectID Type Character Length 8 Format NONE Label LOGIC subject ID

Variable Number 12 Variable AnyLiverTxp Type Numeric Length 8 Format YN. Label Liver transplant

Variable Number 13 Variable RTriSkin Type Numeric Length 8 Format 8.1 Label Right triceps skinfold thickness (mm)

Variable Number 14 Variable RArmSizeCm Type Numeric Length 8 Format NONE Label Right mid arm circumference (cm)

Variable Number 15 Variable INR Type Numeric Length 8 Format 8.1 Label INR

Variable Number 16 Variable AlbuminGdl Type Numeric Length 8 Format NONE Label Albumin (g/dL)

Variable Number 17 Variable ALTUnitsL Type Numeric Length 8 Format BEST12. Label ALT (U/L)

Variable Number 18 Variable ASTUnitsL Type Numeric Length 8 Format BEST12. Label AST (U/L)

Variable Number 19 Variable GGTPUnitsL Type Numeric Length 8 Format BEST12. Label GGTP (U/L)

Variable Number 20 Variable PlateletsCnt Type Numeric Length 8 Format BEST12. Label Platelets (10/mm)

Variable Number 21 Variable BiliTotalMgdl Type Numeric Length 8 Format NONE Label Total or (unconjugated + conjugated) bilirubin (mg/dL)

Variable Number 22 Variable BiliDirectMgdl Type Numeric Length 8 Format NONE Label Direct or conjugated bilirubin (mg/dL)

Variable Number 23 Variable AgeAtExam Type Numeric Length 8 Format NONE Label Age at visit (years)

Variable Number 24 Variable Visit Type Numeric Length 8 Format VISITFMT. Label Visit

Variable Number 25 Variable AgeAtTxp Type Numeric Length 8 Format NONE Label Age (years) at liver transplant

Variable Number 26 Variable race Type Numeric Length 8 Format RACECATF. Label Race

Variable Number 27 Variable PHT Type Numeric Length 8 Format YN. Label PHT status (on this visit)

Variable Number 28 Variable PHTBaseline Type Numeric Length 8 Format YN. Label PHT on entry

Variable Number 29 Variable TxpEnd Type Numeric Length 8 Format YN. Label Liver transplant/death

Variable Number 30 Variable PHTEnd Type Numeric Length 8 Format YN. Label PHT

Variable Number 31 Variable TxpEndYrs Type Numeric Length 8 Format NONE Label Time to liver transplant/death (years)

Variable Number 32 Variable PHTEndYrs Type Numeric Length 8 Format NONE Label Time to PHT (years)

Variable Number 33 Variable ExamYrs Type Numeric Length 8 Format NONE Label Time to visit (years)

Variable Number 34 Variable NeoCholestasis Type Numeric Length 8 Format YN. Label Neonatal cholestasis

Variable Number 35 Variable RespiratoryProblems Type Numeric Length 8 Format YN. Label Respiratory problems

### Data Set Name: [algsbl.sas7bdat](#)

Variable Number 1 Variable agemos Type Numeric Length 8 Format NONE Label Age at exam (months)

Variable Number 2 Variable sex Type Numeric Length 3 Format SEXFMT. Label Sex

Variable Number 3 Variable bmi Type Numeric Length 8 Format NONE Label BMI

Variable Number 4 Variable WeightZScore Type Numeric Length 8 Format NONE Label weight-for-age Z

Variable Number 5 Variable BMIZScore Type Numeric Length 8 Format NONE Label BMI-for-age Z

Variable Number 6 Variable HeightZScore Type Numeric Length 8 Format NONE Label height-for-age Z

Variable Number 7 Variable Ethnicity Type Numeric Length 8 Format ETHNICITYFMT. Label Ethnicity

Variable Number 8 Variable SubjectID Type Character Length 8 Format NONE Label LOGIC subject ID

Variable Number 9 Variable Race Type Numeric Length 8 Format RACEFMT. Label Race

Variable Number 10 Variable IHxKasai Type Numeric Length 8 Format YNMFMT. Label Kasai (initial history)

Variable Number 11 Variable IHxSurgeryDrainAny Type Numeric Length 8 Format YNMFMT. Label Drainage procedure (initial history)

Variable Number 12 Variable IHxALGSPPS Type Numeric Length 8 Format YNMFMT. Label Peripheral pulmonary stenosis

Variable Number 13 Variable IHxALGSPulmStenosis Type Numeric Length 8 Format YNMFMT. Label Pulmonary valve stenosis

Variable Number 14 Variable IHxALGSTetraFallot Type Numeric Length 8 Format YNMFMT. Label Tetralogy of Fallot

Variable Number 15 Variable IHxALGSVentricDef Type Numeric Length 8 Format YNMFMT. Label Ventricular septal defect

Variable Number 16 Variable IHxALGSAtriaDef Type Numeric Length 8 Format YNMFMT. Label Atrial septal defect

Variable Number 17 Variable IHxALGSDeepEyes Type Numeric Length 8 Format YNMFMT. Label Deep-set eyes

Variable Number 18 Variable IHxALGSForehead Type Numeric Length 8 Format YNMFMT. Label Broad forehead

Variable Number 19 Variable IHxALGSChin Type Numeric Length 8 Format YNMFMT. Label Pointed chin

Variable Number 20 Variable IHxALGSPostEmbrotaxon Type Numeric Length 8 Format YNMFMT. Label Posterior embryotoxon

Variable Number 21 Variable IHxALGSAXenfeld Type Numeric Length 8 Format YNMFMT. Label Axenfeld's anomaly

Variable Number 22 Variable IHxALGSButterfly Type Numeric Length 8 Format YNMFMT. Label Butterfly vertebrae

Variable Number 23 Variable IHxALGSDypKidney Type Numeric Length 8 Format YNMFMT. Label Dysplastic kidney

Variable Number 24 Variable IHxALGSSingleKidney Type Numeric Length 8 Format YNMFMT. Label Single kidney

Variable Number 25 Variable IHxALGSRTA Type Numeric Length 8 Format YNMFMT. Label Renal tubular acidosis

Variable Number 26 Variable IHxALGSHeartOther Type Numeric Length 8 Format YNMFMT. Label Other cardiac anomaly

Variable Number 27 Variable IHxALGSEyesOther Type Numeric Length 8 Format YNMFMT. Label Other eye condition

Variable Number 28 Variable IHxALGSSkelOther Type Numeric Length 8 Format YNMFMT. Label Other skeletal condition

Variable Number 29 Variable IHxALGSRenalOther Type Numeric Length 8 Format YNMFMT. Label Other renal structural anomaly

Variable Number 30 Variable IHxAnyRenalAbnormal Type Numeric Length 8 Format YNMFMT. Label Renal abnormality

Variable Number 31 Variable DrainAny Type Numeric Length 8 Format YNMFMT. Label Biliary drainage procedure

Variable Number 32 Variable DrainKasai Type Numeric Length 8 Format YNMFMT. Label Kasai

Variable Number 33 Variable SpleenSize Type Numeric Length 8 Format NONE Label Spleen size (cm) below costal margin (set to 0 if not palpable)

Variable Number 34 Variable HeightCm Type Numeric Length 8 Format NONE Label Height (cm)

Variable Number 35 Variable WeightKg Type Numeric Length 8 Format NONE Label Weight (kg)

Variable Number 36 Variable Pruritus Type Numeric Length 8 Format PRURITUSFMT. Label Pruritus

Variable Number 37 Variable Xanthoma Type Numeric Length 8 Format XANTHOMAFMT. Label Xanthoma

Variable Number 38 Variable INR Type Numeric Length 8 Format NONE Label INR

Variable Number 39 Variable AlbuminGdl Type Numeric Length 8 Format NONE Label Albumin (g/dL)

Variable Number 40 Variable SerumBileAcidsUmolL Type Numeric Length 8 Format NONE Label Total serum bile acids (umol/L)

Variable Number 41 Variable CholesterolMgdl Type Numeric Length 8 Format NONE Label Total cholesterol (mg/dL)

Variable Number 42 Variable VitaminAUgdl Type Numeric Length 8 Format NONE Label Vitamin A (ug/dL)

Variable Number 43 Variable VitaminD125OH2Pgml Type Numeric Length 8 Format NONE Label 1,25 OH2 Vitamin D (pg/mL)

Variable Number 44 Variable VitaminD250HNgml Type Numeric Length 8 Format NONE Label Vitamin D (D25OH) (ng/mL)

Variable Number 45 Variable VitaminEUgdl Type Numeric Length 8 Format NONE Label Vitamin E (ug/mL)

Variable Number 46 Variable ALTUnitsL Type Numeric Length 8 Format NONE Label ALT (U/L)

Variable Number 47 Variable ASTUnitsL Type Numeric Length 8 Format NONE Label AST (U/L)

Variable Number 48 Variable GGTPUnitsL Type Numeric Length 8 Format NONE Label GGTP (U/L)

Variable Number 49 Variable PlateletsCnt Type Numeric Length 8 Format NONE Label Platelets (10/mm)

Variable Number 50 Variable BiliTotalMgdl Type Numeric Length 8 Format NONE Label Total or (unconjugated + conjugated) bilirubin (mg/dL)

Variable Number 51 Variable Baseline Type Numeric Length 8 Format YNMFMT. Label Baseline

Variable Number 52 Variable AgeAtExam Type Numeric Length 8 Format NONE Label Age at exam (years)

Variable Number 53 Variable TimeFromSymptomOnset Type Numeric Length 8 Format NONE Label Time (years) from onset of first disease symptoms

Variable Number 54 Variable Visit Type Numeric Length 8 Format VISITFMT. Label Study visit

Variable Number 55 Variable AgeAtTxp Type Numeric Length 8 Format NONE Label Age (years) at liver transplant

Variable Number 56 Variable AgeAtBaseline Type Numeric Length 8 Format NONE Label Age (years) at baseline LOGIC visit

Variable Number 57 Variable JAG1 Type Numeric Length 8 Format YNMFMT. Label JAG1 mutation

Variable Number 58 Variable NOTCH2 Type Numeric Length 8 Format YNMFMT. Label NOTCH2 mutation

Variable Number 59 Variable ATP8B1 Type Numeric Length 8 Format YNMFMT. Label ATP8B1 mutation

Variable Number 60 Variable ABCB11 Type Numeric Length 8 Format YNMFMT. Label ABCB11 mutation

Variable Number 61 Variable ABCB4 Type Numeric Length 8 Format YNMFMT. Label ABCB4 mutation

Variable Number 62 Variable JAG1DiseaseClass1 Type Numeric Length 8 Format MUTATIONCLASSFMT. Label JAG1 mutation classification

Variable Number 63 Variable NOTCH2DiseaseClass1 Type Numeric Length 8 Format MUTATIONCLASSFMT. Label NOTCH2 mutation classification

Variable Number 64 Variable ALGSMutation Type Character Length 35 Format NONE Label ALGS mutation

Variable Number 65 Variable One Type Numeric Length 8 Format NONE Label Variable with value of 1

Variable Number 66 Variable WholeAgeAtExam Type Numeric Length 8 Format NONE Label Age at exam (years - integer portion)

Variable Number 67 Variable Log10Bili Type Numeric Length 8 Format NONE Label log 10 (bilirubin)

Variable Number 68 Variable Log10ALT Type Numeric Length 8 Format NONE Label log 10 (ALT)

Variable Number 69 Variable Log10AST Type Numeric Length 8 Format NONE Label log 10 (AST)

Variable Number 70 Variable Log10GGTP Type Numeric Length 8 Format NONE Label log 10 (GGTP)

Variable Number 71 Variable Log10Cholesterol Type Numeric Length 8 Format NONE Label log 10 (Cholesterol)

Variable Number 72 Variable APRI Type Numeric Length 8 Format NONE Label AST to platelet ratio index

Variable Number 73 Variable SevereHeartDefect Type Numeric Length 8 Format YNMFMT. Label Severe heart defect

Variable Number 74 Variable HeartDefectCat Type Numeric Length 8 Format HEARTDEFECTCATFMT. Label Heart defect category

Variable Number 75 Variable ALGSMutationClass Type Numeric Length 8 Format MUTATIONCLASSFMT. Label ALGS mutation classification

Variable Number 76 Variable T1 Type Numeric Length 8 Format NONE Label Days from baseline

Variable Number 77 Variable T1Years Type Numeric Length 8 Format NONE Label Years from baseline

Variable Number 78 Variable TestDone Type Numeric Length 8 Format YNMFMT. Label Audiology testing performed

Variable Number 79 Variable TympDone Type Numeric Length 8 Format TYMPFMT. Label Was tympanometry done?

Variable Number 80 Variable TympRE Type Numeric Length 8 Format PFFMT. Label Typanometry right ear

Variable Number 81 Variable TympREFR Type Numeric Length 8 Format FAILRSNFMT. Label Right ear, reason for failure

Variable Number 82 Variable TympREFRText Type Character Length 50 Format \$50. Label Right ear, reason for failure text

Variable Number 83 Variable TympLE Type Numeric Length 8 Format PFFMT. Label Typanometry left ear

Variable Number 84 Variable TympLEFR Type Numeric Length 8 Format FAILRSNFMT. Label Left ear, reason for failure

Variable Number 85 Variable TympLEFRText Type Character Length 50 Format \$50. Label Left ear, reason for failure text

Variable Number 86 Variable SurvTmTxp Type Numeric Length 8 Format NONE Label Survival time from baseline to transplant or death (days)

Variable Number 87 Variable SurvTmTxpYrs Type Numeric Length 8 Format NONE Label Survival time from baseline to transplant or death (years)

Variable Number 88 Variable EventTxp Type Numeric Length 8 Format NONE Label Transplant event (1=transplanted 0=censored)

Variable Number 89 Variable SurvTmPat Type Numeric Length 8 Format NONE Label Survival time from baseline to death (days)

Variable Number 90 Variable SurvTmPatYrs Type Numeric Length 8 Format NONE Label Survival time from baseline to death (years)

Variable Number 91 Variable EventDeath Type Numeric Length 8 Format NONE Label Death event (1=died 0=censored)

Variable Number 92 Variable EventTxpDeath Type Numeric Length 8 Format NONE Label Transplant or death event (1=died or transplanted 0=censored)

Variable Number 93 Variable SurvAgeTxp Type Numeric Length 8 Format NONE Label Survival time from birth to transplant or death (days)

Variable Number 94 Variable SurvAgeTxpYrs Type Numeric Length 8 Format NONE Label Survival time from birth to transplant or death (years)

Variable Number 95 Variable T1AgeYrs Type Numeric Length 8 Format NONE Label Age at entry to risk set (years)

Variable Number 96 Variable SurvAgeTxp5Yrs Type Numeric Length 8 Format NONE Label Survival time from birth to transplant or death (capped at 5 years)

Variable Number 97 Variable EventTxpDeathLe5Yrs Type Numeric Length 8 Format NONE Label Transplant or death event by age 5 (1=died or transplanted 0=censored)

Variable Number 98 Variable AscitesStatus Type Numeric Length 8 Format OUTCOMESTATUSFMT. Label Ascites status

Variable Number 99 Variable AscitesAge Type Numeric Length 8 Format NONE Label Age (years) at ascites

Variable Number 100 Variable GIBleedStatus Type Numeric Length 8 Format OUTCOMESTATUSFMT. Label GI bleed status

Variable Number 101 Variable GIBleedAge Type Numeric Length 8 Format NONE Label Age (years) at GI bleed

Variable Number 102 Variable PPHStatus Type Numeric Length 8 Format OUTCOMESTATUSFMT. Label PPH status

Variable Number 103 Variable PPHAge Type Numeric Length 8 Format NONE Label Age (years) at PPH

Variable Number 104 Variable HPSStatus Type Numeric Length 8 Format OUTCOMESTATUSFMT. Label HPS status

Variable Number 105 Variable HPSAge Type Numeric Length 8 Format NONE Label Age (years) at HPS

Variable Number 106 Variable SplenomegalyStatus Type Numeric Length 8 Format OUTCOMESTATUSFMT. Label Splenomegaly status

Variable Number 107 Variable SplenomegalyAge Type Numeric Length 8 Format NONE Label Age (years) at splenomegaly

Variable Number 108 Variable ThrombocytopeniaStatus Type Numeric Length 8 Format OUTCOMESTATUSFMT. Label Thrombocytopenia status

Variable Number 109 Variable ThrombocytopeniaAge Type Numeric Length 8 Format NONE Label Age at thrombocytopenia

Variable Number 110 Variable BoneFractureStatus Type Numeric Length 8 Format OUTCOMESTATUSFMT. Label Bone fracture status

Variable Number 111 Variable BoneFractureAge Type Numeric Length 8 Format NONE Label Age (years) at bone fracture

Variable Number 112 Variable PartBilDiversion Type Character Length 22 Format NONE Label Partial biliary diversion

Variable Number 113 Variable IlealEx Type Character Length 22 Format NONE Label Ileal exclusion



Variable Number 114 Variable AgeAtBaselineDisplay Type Numeric Length 8 Format NONE Label Age at baseline (years)

Variable Number 115 Variable HeightZScoreDisplay Type Numeric Length 8 Format NONE Label Height Z score

Variable Number 116 Variable WeightZScoreDisplay Type Numeric Length 8 Format NONE Label Weight Z score

Variable Number 117 Variable BMIZScoreDisplay Type Numeric Length 8 Format NONE Label BMI Z score

Variable Number 118 Variable ConductionAllPass Type Numeric Length 8 Format YNMFMT. Label Passed all conduction tests

Variable Number 119 Variable ConductionOneFail Type Numeric Length 8 Format YNMFMT. Label Failed at least one conduction test

Variable Number 120 Variable TxpCompNone Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - none

Variable Number 121 Variable TxpCompFailThrive Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - failure to thrive

Variable Number 122 Variable TxpCompAscites Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - ascites

Variable Number 123 Variable TxpCompCholangitis Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - cholangitis

Variable Number 124 Variable TxpCompCoagulopathy Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - coagulopathy

Variable Number 125 Variable TxpCompVarices Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - varices

Variable Number 126 Variable TxpCompGIBleed Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - GI bleed

Variable Number 127 Variable TxpCompEncephalopathy Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - encephalopathy

Variable Number 128 Variable TxpCompHepatopulmonary Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - hepatopulmonary syndrome

Variable Number 129 Variable TxpCompHepatorenal Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - hepatorenal syndrome

Variable Number 130 Variable TxpCompFracture Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - bone fracture

Variable Number 131 Variable TxpCompXanthomatosis Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - xanthomatosis

Variable Number 132 Variable TxpCompPruritus Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - pruritus

Variable Number 133 Variable TxpCompOther Type Numeric Length 8 Format YNMFMT. Label Complications noted at transplant - other

Variable Number 134 Variable TxpCompOSTXT Type Character Length 200 Format NONE Label Complications noted at transplant - other text

Variable Number 135 Variable DeathCauseText Type Character Length 200 Format NONE Label Cause of death

Variable Number 136 Variable MinBaselineDt Type Numeric Length 8 Format MMDDYY10. Label Min baseline

Variable Number 137 Variable MaxBaselineDt Type Numeric Length 8 Format MMDDYY10. Label Max baseline

Variable Number 138 Variable MaxFuDt Type Numeric Length 8 Format MMDDYY10. Label Max follow-up date

Variable Number 139 Variable MaxFuAge Type Numeric Length 8 Format NONE Label Oldest follow up age

## Data Set Name: [algsformats.sas7bdat](#)

Variable Number 1 Variable FMTNAME Type Character Length 32 Format . Label Format name

Variable Number 2 Variable START Type Character Length 16 Format . Label Starting value for format

Variable Number 3 Variable END Type Character Length 16 Format . Label Ending value for format

Variable Number 4 Variable LABEL Type Character Length 113 Format . Label Format value label

Variable Number 5 Variable MIN Type Numeric Length 3 Format . Label Minimum length

Variable Number 6 Variable MAX Type Numeric Length 3 Format . Label Maximum length

Variable Number 7 Variable DEFAULT Type Numeric Length 3 Format . Label Default length

Variable Number 8 Variable LENGTH Type Numeric Length 3 Format . Label Format length

Variable Number 9 Variable FUZZ Type Numeric Length 8 Format . Label Fuzz value

Variable Number 10 Variable PREFIX Type Character Length 2 Format . Label Prefix characters

Variable Number 11 Variable MULT Type Numeric Length 8 Format . Label Multiplier

Variable Number 12 Variable FILL Type Character Length 1 Format . Label Fill character

Variable Number 13 Variable NOEDIT Type Numeric Length 3 Format . Label Is picture string noedit?

Variable Number 14 Variable TYPE Type Character Length 1 Format . Label Type of format  
Variable Number 15 Variable SEXCL Type Character Length 1 Format . Label Start exclusion  
Variable Number 16 Variable EEXCL Type Character Length 1 Format . Label End exclusion  
Variable Number 17 Variable HLO Type Character Length 13 Format . Label Additional information  
Variable Number 18 Variable DECSEP Type Character Length 1 Format . Label Decimal separator  
Variable Number 19 Variable DIG3SEP Type Character Length 1 Format . Label Three-digit separator  
Variable Number 20 Variable DATATYPE Type Character Length 8 Format . Label Date/time/datetime?  
Variable Number 21 Variable LANGUAGE Type Character Length 8 Format . Label Language for date strings

### Data Set Name: [algsfu.sas7bdat](#)

Variable Number 1 Variable agemos Type Numeric Length 8 Format NONE Label Age at exam (months)  
Variable Number 2 Variable sex Type Numeric Length 3 Format SEXFMT. Label Sex  
Variable Number 3 Variable bmi Type Numeric Length 8 Format NONE Label BMI  
Variable Number 4 Variable WeightZScore Type Numeric Length 8 Format NONE Label weight-for-age Z  
Variable Number 5 Variable BMIZScore Type Numeric Length 8 Format NONE Label BMI-for-age Z  
Variable Number 6 Variable HeightZScore Type Numeric Length 8 Format NONE Label height-for-age Z  
Variable Number 7 Variable Ethnicity Type Numeric Length 8 Format ETHNICITYFMT. Label Ethnicity  
Variable Number 8 Variable SubjectID Type Character Length 8 Format NONE Label LOGIC subject ID  
Variable Number 9 Variable Race Type Numeric Length 8 Format RACEFMT. Label Race  
Variable Number 10 Variable IHxKasai Type Numeric Length 8 Format YNMFMT. Label Kasai (initial history)  
Variable Number 11 Variable IHxSurgeryDrainAny Type Numeric Length 8 Format YNMFMT. Label Drainage procedure (initial history)  
Variable Number 12 Variable IHxALGSPPS Type Numeric Length 8 Format YNMFMT. Label Peripheral pulmonary stenosis  
Variable Number 13 Variable IHxALGSPulmStenosis Type Numeric Length 8 Format YNMFMT. Label Pulmonary valve stenosis  
Variable Number 14 Variable IHxALGSTetraFallot Type Numeric Length 8 Format YNMFMT. Label Tetralogy of Fallot  
Variable Number 15 Variable IHxALGSVentricDef Type Numeric Length 8 Format YNMFMT. Label Ventricular septal defect

Variable Number 16 Variable IHxALGSAtriaDef Type Numeric Length 8 Format YNMFMT. Label Atrial septal defect

Variable Number 17 Variable IHxALGSDeepEyes Type Numeric Length 8 Format YNMFMT. Label Deep-set eyes

Variable Number 18 Variable IHxALGSForehead Type Numeric Length 8 Format YNMFMT. Label Broad forehead

Variable Number 19 Variable IHxALGSChin Type Numeric Length 8 Format YNMFMT. Label Pointed chin

Variable Number 20 Variable IHxALGSPostEmbrotaxon Type Numeric Length 8 Format YNMFMT. Label Posterior embryotoxon

Variable Number 21 Variable IHxALGSAXenfeld Type Numeric Length 8 Format YNMFMT. Label Axenfeld's anomaly

Variable Number 22 Variable IHxALGSButterfly Type Numeric Length 8 Format YNMFMT. Label Butterfly vertebrae

Variable Number 23 Variable IHxALGSDypKidney Type Numeric Length 8 Format YNMFMT. Label Dysplastic kidney

Variable Number 24 Variable IHxALGSSingleKidney Type Numeric Length 8 Format YNMFMT. Label Single kidney

Variable Number 25 Variable IHxALGSRTA Type Numeric Length 8 Format YNMFMT. Label Renal tubular acidosis

Variable Number 26 Variable IHxALGSHeartOther Type Numeric Length 8 Format YNMFMT. Label Other cardiac anomaly

Variable Number 27 Variable IHxALGSEyesOther Type Numeric Length 8 Format YNMFMT. Label Other eye condition

Variable Number 28 Variable IHxALGSSkelOther Type Numeric Length 8 Format YNMFMT. Label Other skeletal condition

Variable Number 29 Variable IHxALGSRenalOther Type Numeric Length 8 Format YNMFMT. Label Other renal structural anomaly

Variable Number 30 Variable IHxAnyRenalAbnormal Type Numeric Length 8 Format YNMFMT. Label Renal abnormality

Variable Number 31 Variable DrainAny Type Numeric Length 8 Format YNMFMT. Label Biliary drainage procedure

Variable Number 32 Variable DrainKasai Type Numeric Length 8 Format YNMFMT. Label Kasai

Variable Number 33 Variable SpleenSize Type Numeric Length 8 Format NONE Label Spleen size (cm) below costal margin (set to 0 if not palpable)

Variable Number 34 Variable HeightCm Type Numeric Length 8 Format NONE Label Height (cm)

Variable Number 35 Variable WeightKg Type Numeric Length 8 Format NONE Label Weight (kg)

Variable Number 36 Variable Pruritus Type Numeric Length 8 Format PRURITUSFMT. Label Pruritus

Variable Number 37 Variable Xanthoma Type Numeric Length 8 Format XANTHOMAFMT. Label Xanthoma

Variable Number 38 Variable INR Type Numeric Length 8 Format NONE Label INR

Variable Number 39 Variable AlbuminGdl Type Numeric Length 8 Format NONE Label Albumin (g/dL)

Variable Number 40 Variable SerumBileAcidsUmolL Type Numeric Length 8 Format NONE Label Total serum bile acids (umol/L)

Variable Number 41 Variable CholesterolMgdl Type Numeric Length 8 Format NONE Label Total cholesterol (mg/dL)

Variable Number 42 Variable VitaminAUgdl Type Numeric Length 8 Format NONE Label Vitamin A (ug/dL)

Variable Number 43 Variable VitaminD125OH2Pgml Type Numeric Length 8 Format NONE Label 1,25 OH2 Vitamin D (pg/mL)

Variable Number 44 Variable VitaminD250HNgml Type Numeric Length 8 Format NONE Label Vitamin D (D25OH) (ng/mL)

Variable Number 45 Variable VitaminEUgdl Type Numeric Length 8 Format NONE Label Vitamin E (ug/mL)

Variable Number 46 Variable ALTUnitsL Type Numeric Length 8 Format NONE Label ALT (U/L)

Variable Number 47 Variable ASTUnitsL Type Numeric Length 8 Format NONE Label AST (U/L)

Variable Number 48 Variable GGTPUnitsL Type Numeric Length 8 Format NONE Label GGTP (U/L)

Variable Number 49 Variable PlateletsCnt Type Numeric Length 8 Format NONE Label Platelets (10/mm)

Variable Number 50 Variable BiliTotalMgdl Type Numeric Length 8 Format NONE Label Total or (unconjugated + conjugated) bilirubin (mg/dL)

Variable Number 51 Variable Baseline Type Numeric Length 8 Format YNMFMT. Label Baseline

Variable Number 52 Variable AgeAtExam Type Numeric Length 8 Format NONE Label Age at exam (years)

Variable Number 53 Variable TimeFromSymptomOnset Type Numeric Length 8 Format NONE Label Time (years) from onset of first disease symptoms

Variable Number 54 Variable Visit Type Numeric Length 8 Format VISITFMT. Label Study visit

Variable Number 55 Variable AgeAtTxp Type Numeric Length 8 Format NONE Label Age (years) at liver transplant

Variable Number 56 Variable AgeAtBaseline Type Numeric Length 8 Format NONE Label Age (years) at baseline LOGIC visit

Variable Number 57 Variable JAG1 Type Numeric Length 8 Format YNMFMT. Label JAG1 mutation

Variable Number 58 Variable NOTCH2 Type Numeric Length 8 Format YNMFMT. Label NOTCH2 mutation

Variable Number 59 Variable ATP8B1 Type Numeric Length 8 Format YNMFMT. Label ATP8B1 mutation

Variable Number 60 Variable ABCB11 Type Numeric Length 8 Format YNMFMT. Label ABCB11 mutation

Variable Number 61 Variable ABCB4 Type Numeric Length 8 Format YNMFMT. Label ABCB4 mutation

Variable Number 62 Variable JAG1DiseaseClass1 Type Numeric Length 8 Format MUTATIONCLASSFMT. Label JAG1 mutation classification

Variable Number 63 Variable NOTCH2DiseaseClass1 Type Numeric Length 8 Format MUTATIONCLASSFMT. Label NOTCH2 mutation classification

Variable Number 64 Variable ALGSMutation Type Character Length 35 Format NONE Label ALGS mutation

Variable Number 65 Variable One Type Numeric Length 8 Format NONE Label Variable with value of 1

Variable Number 66 Variable WholeAgeAtExam Type Numeric Length 8 Format NONE Label Age at exam (years - integer portion)

Variable Number 67 Variable Log10Bili Type Numeric Length 8 Format NONE Label log 10 (bilirubin)

Variable Number 68 Variable Log10ALT Type Numeric Length 8 Format NONE Label log 10 (ALT)

Variable Number 69 Variable Log10AST Type Numeric Length 8 Format NONE Label log 10 (AST)

Variable Number 70 Variable Log10GGTP Type Numeric Length 8 Format NONE Label log 10 (GGTP)

Variable Number 71 Variable Log10Cholesterol Type Numeric Length 8 Format NONE Label log 10 (Cholesterol)

Variable Number 72 Variable APRI Type Numeric Length 8 Format NONE Label AST to platelet ratio index

Variable Number 73 Variable SevereHeartDefect Type Numeric Length 8 Format YNMFMT. Label Severe heart defect

Variable Number 74 Variable HeartDefectCat Type Numeric Length 8 Format HEARTDEFECTCATFMT. Label Heart defect category

Variable Number 75 Variable ALGSMutationClass Type Numeric Length 8 Format MUTATIONCLASSFMT. Label ALGS mutation classification

Variable Number 76 Variable T1 Type Numeric Length 8 Format NONE Label Days from baseline

Variable Number 77 Variable T1Years Type Numeric Length 8 Format NONE Label Years from baseline

## Data Set Name: `algsnhcontrolsbl.sas7bdat`

Variable Number 1 Variable `ParticipantID` Type Character Length 25 Format \$25. Label `ChiLDReN` participant ID

Variable Number 2 Variable `SampWeight` Type Numeric Length 8 Format NONE Label Sample weight (based on age distribution)

Variable Number 3 Variable `AgeAtBaseline` Type Numeric Length 8 Format NONE Label Age at Baseline (years)

Variable Number 4 Variable `AgeGrp` Type Numeric Length 8 Format `AGEGRPFMT`. Label Age group

Variable Number 5 Variable `Gender` Type Numeric Length 8 Format `SEXFMT`. Label Sex

Variable Number 6 Variable `Race` Type Numeric Length 8 Format `RACEFMT`. Label Race

Variable Number 7 Variable `Ethnicity` Type Numeric Length 8 Format `ETHNICITYFMT`. Label Ethnicity

Variable Number 8 Variable `CSS` Type Numeric Length 8 Format `CSSFMT`. Label Clinician scratch scale

Variable Number 9 Variable `PedsQLTotalParent` Type Numeric Length 8 Format NONE Label PedsQL total score

Variable Number 10 Variable `BiliTotalMgdl` Type Numeric Length 8 Format NONE Label Total bilirubin (mg/dl)

Variable Number 11 Variable `CholesterolMgdl` Type Numeric Length 8 Format NONE Label Cholesterol (mg/dl)

Variable Number 12 Variable `SerumBileAcidsUmolL` Type Numeric Length 8 Format NONE Label Serum bile acids (umol/L)

Variable Number 13 Variable `ASTUnitsL` Type Numeric Length 8 Format NONE Label AST

Variable Number 14 Variable `ALTUnitsL` Type Numeric Length 8 Format NONE Label ALT

Variable Number 15 Variable `GGTPUnitsL` Type Numeric Length 8 Format NONE Label GGTP

Variable Number 16 Variable `AlbuminGdl` Type Numeric Length 8 Format NONE Label Albumin (g/dl)

Variable Number 17 Variable `PlateletsCnt` Type Numeric Length 8 Format NONE Label Platelet count (10/mm)

Variable Number 18 Variable `HeightZScore` Type Numeric Length 8 Format NONE Label Height Z-score

Variable Number 19 Variable `WeightZScore` Type Numeric Length 8 Format NONE Label Weight Z-score

Variable Number 20 Variable `SurvTmTxpDeathYrs` Type Numeric Length 8 Format NONE Label Survival time from baseline to transplant or death (years)

Variable Number 21 Variable `SurvTmCEAscitesYrs` Type Numeric Length 8 Format NONE Label Survival time from baseline to ascites (years)

Variable Number 22 Variable SurvAgeTxpDeathYrs Type Numeric Length 8 Format NONE Label Survival time from birth to transplant or death (years)

Variable Number 23 Variable EventDeath Type Numeric Length 8 Format NONE Label Death event (1=died 0=censored)

Variable Number 24 Variable EventTxp Type Numeric Length 8 Format NONE Label Transplant event (1=transplanted 0=censored)

Variable Number 25 Variable EventTxpDeath Type Numeric Length 8 Format NONE Label Transplant or death event (1=died or transplanted 0=censored)

Variable Number 26 Variable EventCEAscites Type Numeric Length 8 Format NONE Label Clinically evident ascites (1=event 0=censored)

Variable Number 27 Variable EventCEVaricealBleed Type Numeric Length 8 Format NONE Label Clinically evident variceal hemorrhage (1=event 0=censored)

Variable Number 28 Variable EventDrain Type Numeric Length 8 Format NONE Label Partial biliary diversion or ileal exclusion (1=event 0=censored)

Variable Number 29 Variable EventComposite Type Numeric Length 8 Format NONE Label Earliest of transplant, death, ascites, bleed (1=event 0=censored)

Variable Number 30 Variable CauseOfDeath Type Character Length 4000 Format \$4000. Label Cause of death

## Data Set Name: [algsnhcontrolsfu.sas7bdat](#)

Variable Number 1 Variable ParticipantID Type Character Length 25 Format \$25. Label ChiLDReN participant ID

Variable Number 2 Variable SampWeight Type Numeric Length 8 Format NONE Label Sample weight (based on age distribution)

Variable Number 3 Variable AgeAtBaseline Type Numeric Length 8 Format NONE Label Age at Baseline (years)

Variable Number 4 Variable AgeAtExam Type Numeric Length 8 Format NONE Label Age at Exam (years)

Variable Number 5 Variable Visit Type Numeric Length 8 Format VISITFMT. Label Study visit

Variable Number 6 Variable ExamTimeYrs Type Numeric Length 8 Format NONE Label Time (years) from baseline to exam

Variable Number 7 Variable TransplantTimeYrs Type Numeric Length 8 Format NONE Label Time (years) from baseline to transplant

Variable Number 8 Variable DeathTimeYrs Type Numeric Length 8 Format NONE Label Time (years) from baseline to death DrainPartBilDiversionTimeYrsTime (years) from baseline to partial biliary diversion

Variable Number 9 Variable AgeGrp Type Numeric Length 8 Format AGEGRPFMT. Label Age group



Variable Number 10 Variable Gender Type Numeric Length 8 Format SEXFMT. Label Sex

Variable Number 11 Variable Race Type Numeric Length 8 Format RACEFMT. Label Race

Variable Number 12 Variable Ethnicity Type Numeric Length 8 Format ETHNICITYFMT. Label Ethnicity

Variable Number 13 Variable CSS Type Numeric Length 8 Format CSSFMT. Label Clinician scratch scale

Variable Number 14 Variable BiliTotalMgdl Type Numeric Length 8 Format NONE Label Total bilirubin (mg/dl)

Variable Number 15 Variable ALTUnitsL Type Numeric Length 8 Format NONE Label ALT

Variable Number 16 Variable AlbuminGdl Type Numeric Length 8 Format NONE Label Albumin (g/dl)

Variable Number 17 Variable PlateletsCnt Type Numeric Length 8 Format NONE Label Platelet count (10/mm)

Variable Number 18 Variable HeightZScore Type Numeric Length 8 Format NONE Label Height Z-score

Variable Number 19 Variable WeightZScore Type Numeric Length 8 Format NONE Label Weight Z-score

Variable Number 20 Variable CSS\_BL Type Numeric Length 8 Format NONE Label CSS at baseline

Variable Number 21 Variable BiliTotal\_BL Type Numeric Length 8 Format NONE Label Total bilirubin at baseline

Variable Number 22 Variable ALT\_BL Type Numeric Length 8 Format NONE Label ALT at baseline

Variable Number 23 Variable Albumin\_BL Type Numeric Length 8 Format NONE Label Albumin at baseline

Variable Number 24 Variable Platelet\_BL Type Numeric Length 8 Format NONE Label Platelet count at baseline

Variable Number 25 Variable HeightZ\_BL Type Numeric Length 8 Format NONE Label Height Z-score at baseline

Variable Number 26 Variable WeightZ\_BL Type Numeric Length 8 Format NONE Label Weight Z-score at baseline

Variable Number 27 Variable CSS\_Change Type Numeric Length 8 Format NONE Label CSS change from baseline

Variable Number 28 Variable BiliTotal\_Change Type Numeric Length 8 Format NONE Label Total bilirubin change from baseline

Variable Number 29 Variable ALT\_Change Type Numeric Length 8 Format NONE Label ALT change from baseline

Variable Number 30 Variable Albumin\_Change Type Numeric Length 8 Format NONE Label Albumin change from baseline

Variable Number 31 Variable Platelet\_Change Type Numeric Length 8 Format NONE Label Platelet count change from baseline

Variable Number 32 Variable HeightZ\_Change Type Numeric Length 8 Format NONE Label Height Z-score change from baseline

Variable Number 33 Variable WeightZ\_Change Type Numeric Length 8 Format NONE Label Weight Z-score change from baseline

Variable Number 34 Variable CSS\_ClinSig Type Numeric Length 8 Format NONE Label Clinically significant change in CSS

Variable Number 35 Variable BiliTotal\_ClinSig Type Numeric Length 8 Format NONE Label Clinically significant change in total bilirubin

Variable Number 36 Variable ALT\_ClinSig Type Numeric Length 8 Format NONE Label Clinically significant change in ALT

Variable Number 37 Variable Platelet\_ClinSig Type Numeric Length 8 Format NONE Label Clinically significant change in platelet count

Variable Number 38 Variable Albumin\_ClinSig Type Numeric Length 8 Format NONE Label Clinically significant change in Alubmin

Variable Number 39 Variable WeightZ\_ClinSig Type Numeric Length 8 Format NONE Label Clinically significant change in weight Z-score

Variable Number 40 Variable HeightZ\_ClinSig Type Numeric Length 8 Format NONE Label Clinically significant change in height Z-score

Variable Number 41 Variable Urso\_Flag Type Numeric Length 8 Format NONE Label Ursodeoxycholic acid use noted (1=yes, 0=no)

Variable Number 42 Variable Rifampin\_Flag Type Numeric Length 8 Format NONE Label Rifampin use noted (1=yes, 0=no)

Variable Number 43 Variable Antihist\_Flag Type Numeric Length 8 Format NONE Label Antihistamine use noted (1=yes, 0=no)

Variable Number 44 Variable Cholestyryra\_Flag Type Numeric Length 8 Format NONE Label Cholestyramine use noted (1=yes, 0=no)

Variable Number 45 Variable Phenobarb\_Flag Type Numeric Length 8 Format NONE Label Phenobarbital use noted (1=yes, 0=no)

Variable Number 46 Variable NonOralNutrition\_Flag Type Numeric Length 8 Format NONE Label Non-oral nutrition use noted (1=yes, 0=no)

Variable Number 47 Variable Naltrexone\_Flag Type Numeric Length 8 Format NONE Label Naltrexone use noted (1=yes, 0=no)

Variable Number 48 Variable DrainPartBilDiversionTimeYrs Type Numeric Length 8 Format NONE Label NONE

Variable Number 49 Variable DrainIlealExTimeYrs Type Numeric Length 8 Format NONE Label Time (years) from baseline to ileal exclusion

## Data Set Name: [algsnhcontrolselect.sas7bdat](#)

Variable Number 1 Variable ParticipantID Type Character Length 25 Format \$25. Label ChiLDReN participant ID

Variable Number 2 Variable Diagnosis Type Numeric Length 8 Format DIAGNOSISFMT. Label LOGIC diagnosis

Variable Number 3 Variable IncludeStudyDate Type Numeric Length 8 Format NONE Label LOGIC baseline before 1/1/2020 (1=yes, 0=no)

Variable Number 4 Variable IncludePreTxp Type Numeric Length 8 Format NONE Label Pre-transplant enrollment flag (1=Pre-transplant, 0=Post-transplant)

Variable Number 5 Variable IncludeDisease Type Numeric Length 8 Format NONE Label Does participant have disease? (1=yes, 0=no (unaffected sibling))

Variable Number 6 Variable IncludeAge Type Numeric Length 8 Format NONE Label Age 1 to 18 years (1=yes, 0=no)

Variable Number 7 Variable StudyAgeInclude Type Numeric Length 8 Format NONE Label Turned 1 year old during LOGIC study (1=yes, 0=no)

Variable Number 8 Variable IncludeAnyFollowUp Type Numeric Length 8 Format NONE Label Has some follow-up information (1=yes, 0=no)

Variable Number 9 Variable IncludeCholestasis Type Numeric Length 8 Format NONE Label Evidence of cholestasis (1=yes, 0=no)

Variable Number 10 Variable Cholestasis1 Type Numeric Length 8 Format NONE Label Serum bile acids > 50 (1=yes, 0=no)

Variable Number 11 Variable Cholestasis2 Type Numeric Length 8 Format NONE Label Direct bilirubin > 1 (1=yes, 0=no)

Variable Number 12 Variable Cholestasis3 Type Numeric Length 8 Format NONE Label Conjugated bilirubin > 1 (1=yes, 0=no)

Variable Number 13 Variable Cholestasis4 Type Numeric Length 8 Format NONE Label GGTP > 150 (1=yes, 0=no)

Variable Number 14 Variable Cholestasis5 Type Numeric Length 8 Format NONE Label CSS 3 or 4 (1=yes, 0=no)

Variable Number 15 Variable IncludeScratch Type Numeric Length 8 Format NONE Label CSS 2 or higher at baseline (1=yes, 0=no)

Variable Number 16 Variable ExcludeDrugTrial Type Numeric Length 8 Format NONE Label Participant in Drug Trial (1=yes, 0=no)

Variable Number 17 Variable ExcludeKasai Type Numeric Length 8 Format NONE Label History of Kasai portoenterostomy (1=yes, 0=no)

Variable Number 18 Variable ExcludeDrain Type Numeric Length 8 Format NONE Label History of biliary diversion or ileal exclusion (1=yes, 0=no)

Variable Number 19 Variable ExcludeOtherLiverDgn Type Numeric Length 8 Format NONE Label Other liver disease noted (1=yes, 0=no)

Variable Number 20 Variable ExcludeALT Type Numeric Length 8 Format NONE Label ALT over 600 (15 x ULN) (1=yes, 0=no)

Variable Number 21 Variable ExcludeAlbumin Type Numeric Length 8 Format NONE Label Albumin under 3 (1=yes, 0=no)

Variable Number 22 Variable ExcludeCEAscites Type Numeric Length 8 Format NONE Label History of clinically evident ascites (1=yes, 0=no)

Variable Number 23 Variable ExcludeGIBleed Type Numeric Length 8 Format NONE Label History of GI bleed (1=yes, 0=no)

Variable Number 24 Variable ExcludeTxpList Type Numeric Length 8 Format NONE Label History of liver transplant waiting list (1=yes, 0=no)

Variable Number 25 Variable IncludeFlag Type Numeric Length 8 Format NONE Label Included in study (1=yes, 0=no)

Variable Number 26 Variable MinConsent Type Numeric Length 8 Format MMDDYY10. Label Earliest eligible LOGIC consent date

Variable Number 27 Variable MaxConsent Type Numeric Length 8 Format MMDDYY10. Label Most recent eligible LOGIC consent date

## Data Set Name: [child\\_univariate.sas7bdat](#)

Variable Number 1 Variable CTXSTDID Type Numeric Length 8 Format . Label De-Identified Subject ID

Variable Number 2 Variable PHYSICAL Type Numeric Length 8 Format . Label QOL Physical

Variable Number 3 Variable PSYCHOSOCIAL Type Numeric Length 8 Format . Label QOL psychosocial

Variable Number 4 Variable TOTAL Type Numeric Length 8 Format . Label QOL Total

Variable Number 5 Variable WAZ Type Numeric Length 8 Format . Label Weight z-scores

Variable Number 6 Variable HAZ Type Numeric Length 8 Format . Label Height z-scores

Variable Number 7 Variable BT\_N Type Numeric Length 8 Format . Label Total Bilirubin (mg/dL)

Variable Number 8 Variable DIS\_GRP Type Character Length 25 Format . Label Disease Group - ALGS, BAD, PFIC, a1-AT

Variable Number 9 Variable GRP Type Numeric Length 8 Format . Label Eligibility group

### Data Set Name: demographics.sas7bdat

Variable Number 1 Variable CTXSTDID Type Numeric Length 8 Format . Label De-Identified Subject ID

Variable Number 2 Variable PHYSICAL Type Numeric Length 8 Format . Label QOL Physical

Variable Number 3 Variable TOTAL Type Numeric Length 8 Format . Label QOL Total

Variable Number 4 Variable WAZ Type Numeric Length 8 Format . Label Weight z-scores

Variable Number 5 Variable HAZ Type Numeric Length 8 Format . Label Height z-scores

Variable Number 6 Variable BT\_N Type Numeric Length 8 Format . Label Total Bilirubin (mg/dL)

Variable Number 7 Variable GGTP\_N Type Numeric Length 8 Format . Label GGTP (U/L)

Variable Number 8 Variable INR\_N Type Numeric Length 8 Format . Label INR

Variable Number 9 Variable WBC\_N Type Numeric Length 8 Format . Label WBC (10/mm)

Variable Number 10 Variable PLATELETS\_N Type Numeric Length 8 Format . Label Platelet (10/mm)

Variable Number 11 Variable ALT\_N Type Numeric Length 8 Format . Label ALT (U/L)

Variable Number 12 Variable ALBUMIN\_N Type Numeric Length 8 Format . Label Albumin (g/dL)

Variable Number 13 Variable RACE Type Character Length 12 Format . Label Race

Variable Number 14 Variable GENDER Type Character Length 6 Format . Label Gender - 1=male  
2=female

Variable Number 15 Variable TX\_LISTING Type Character Length 1 Format . Label Transplant Listing -  
1=Listed for Transplant

Variable Number 16 Variable DIS\_GRP Type Character Length 25 Format . Label Disease Group - ALGS,  
BAD, PFIC, a1-AT

Variable Number 17 Variable GRP Type Numeric Length 8 Format . Label Eligibility group

Variable Number 18 Variable AGE\_AT\_TEST Type Numeric Length 8 Format . Label Age at QOL survey  
(years)

### Data Set Name: dxadata.sas7bdat

Variable Number 1 Variable BMDSPINE Type Numeric Length 8 Format . Label Total spine BMD

Variable Number 2 Variable BMDSPINEZSCORE Type Numeric Length 8 Format . Label Total spine BMD  
Z-score

Variable Number 3 Variable BMDNECK Type Numeric Length 8 Format . Label Hip neck BMD

Variable Number 4 Variable BMDNECKZSCORE Type Numeric Length 8 Format . Label Hip neck BMD Z-score

Variable Number 5 Variable BMDHIP Type Numeric Length 8 Format . Label Total hip BMD

Variable Number 6 Variable BMDHIPZSCORE Type Numeric Length 8 Format . Label Total hip BMD Z-score

Variable Number 7 Variable BMDSPINEWTHTFATZSCORE Type Numeric Length 8 Format . Label Total spine BMD Z-score adjusted for weight, height, fat

Variable Number 8 Variable BMDNECKWTHTFATZSCORE Type Numeric Length 8 Format . Label Hip neck BMD Z-score adjusted for weight, height, fat

Variable Number 9 Variable BMDHIPWTHTFATZSCORE Type Numeric Length 8 Format . Label Total hip BMD Z-score adjusted for weight, height, fat

Variable Number 10 Variable BMCTOTALWTHTFATZSCORE Type Numeric Length 8 Format . Label Whole body BMC Z-score adjusted for weight, height, fat

Variable Number 11 Variable BMDTOTALWTHTFATZSCORE Type Numeric Length 8 Format . Label Whole body BMD Z-score adjusted for weight, height, fat

Variable Number 12 Variable BMCSPINEWTHTFATZSCORE Type Numeric Length 8 Format . Label Total spine BMC Z-score adjusted for weight, height, fat

Variable Number 13 Variable BMDSPINEWTHTZSCORE Type Numeric Length 8 Format . Label Total spine BMD Z-score adjusted for weight, height

Variable Number 14 Variable BMDNECKWTHTZSCORE Type Numeric Length 8 Format . Label Hip neck BMD Z-score adjusted for weight, height

Variable Number 15 Variable BMDHIPWTHTZSCORE Type Numeric Length 8 Format . Label Total hip BMD Z-score adjusted for weight, height

Variable Number 16 Variable BMCTOTALWTHTZSCORE Type Numeric Length 8 Format . Label Whole body BMC Z-score adjusted for weight, height

Variable Number 17 Variable BMDTOTALWTHTZSCORE Type Numeric Length 8 Format . Label Whole body BMD Z-score adjusted for weight, height

Variable Number 18 Variable BMCSPINEWTHTZSCORE Type Numeric Length 8 Format . Label Total spine BMC Z-score adjusted for weight, height

Variable Number 19 Variable MEASUREDPCTLEAN Type Numeric Length 8 Format . Label Measured lean body mass (%)

Variable Number 20 Variable BMCTOTAL Type Numeric Length 8 Format . Label Whole body BMC

Variable Number 21 Variable BMDTOTAL Type Numeric Length 8 Format . Label Whole body BMD

Variable Number 22 Variable BMDSUBTOTAL Type Numeric Length 8 Format . Label Whole body minus head BMD

Variable Number 23 Variable BMCSUBTOTAL Type Numeric Length 8 Format . Label Whole body minus head BMC

Variable Number 24 Variable BMDTOTALZSCORE Type Numeric Length 8 Format . Label Whole body BMD Z-score

Variable Number 25 Variable BMDSUBTOTALZSCORE Type Numeric Length 8 Format . Label Whole body minus head BMD Z-score

Variable Number 26 Variable BMCTOTALZSCORE Type Numeric Length 8 Format . Label Whole body BMC Z-score

Variable Number 27 Variable BMCSUBTOTALZSCORE Type Numeric Length 8 Format . Label Whole body minus head BMC Z-score

Variable Number 28 Variable BMCSUBTOTALWHTHFATZSCORE Type Numeric Length 8 Format . Label Whole body minus head BMC Z-score adjusted for weight, height, fat

Variable Number 29 Variable BMDSUBTOTALWHTHFATZSCORE Type Numeric Length 8 Format . Label Whole body minus head BMD Z-score adjusted for weight, height, fat

Variable Number 30 Variable BMCNECKWHTHFATZSCORE Type Numeric Length 8 Format . Label Hip neck BMC Z-score adjusted for weight, height, fat

Variable Number 31 Variable BMCHIPWHTHFATZSCORE Type Numeric Length 8 Format . Label Total hip BMC Z-score adjusted for weight, height, fat

Variable Number 32 Variable BMCSUBTOTALWHTHZSCORE Type Numeric Length 8 Format . Label Whole body minus head BMC Z-score adjusted for weight, height

Variable Number 33 Variable BMDSUBTOTALWHTHZSCORE Type Numeric Length 8 Format . Label Whole body minus head BMD Z-score adjusted for weight, height

Variable Number 34 Variable BMCNECKWHTHZSCORE Type Numeric Length 8 Format . Label Hip neck BMC Z-score adjusted for weight, height

Variable Number 35 Variable BMCHIPWHTHZSCORE Type Numeric Length 8 Format . Label Total hip BMC Z-score adjusted for weight, height

Variable Number 36 Variable SUBJECTID Type Character Length 7 Format . Label LOGIC subject ID

Variable Number 37 Variable LOGICGROUP Type Numeric Length 8 Format . Label LOGIC study group

Variable Number 38 Variable NODXA Type Numeric Length 8 Format . Label Flag for subjects without a scan used as matched comparison (0=DXA scan, 1=no DXA scan)

Variable Number 39 Variable DGNDTAIL Type Numeric Length 8 Format . Label Detailed diagnosis from diagnosis CRF

Variable Number 40 Variable DIAGNOSIS Type Numeric Length 8 Format . Label Diagnosis group

Variable Number 41 Variable SEX Type Numeric Length 8 Format . Label Gender

Variable Number 42 Variable ETHNICITY Type Numeric Length 8 Format . Label Ethnicity

Variable Number 43 Variable RACE Type Numeric Length 8 Format . Label Race

Variable Number 44 Variable AGEATSCANYEARS Type Numeric Length 8 Format . Label Age (years) at the time of scan

Variable Number 45 Variable AGEATBASELINEYEARS Type Numeric Length 8 Format . Label Age (years) at LOGIC study baseline

Variable Number 46 Variable AGEATSCANORMATCH Type Numeric Length 8 Format . Label Age (years) at DXA scan or date used to match non-DXA subject

Variable Number 47 Variable HEIGHTCM Type Numeric Length 8 Format . Label Height (cm) closest to scan

Variable Number 48 Variable WEIGHTKG Type Numeric Length 8 Format . Label Weight (kg) closest to scan

Variable Number 49 Variable SPLEENSIZE Type Numeric Length 8 Format . Label Spleen size (cm below costal margin)

Variable Number 50 Variable BILITOTAL Type Numeric Length 8 Format . Label Total bilirubin (mg/dL)

Variable Number 51 Variable BILIDIRECT Type Numeric Length 8 Format . Label Direct bilirubin (mg/dL)

Variable Number 52 Variable ALBUMIN Type Numeric Length 8 Format . Label Albumin (g/dL)

Variable Number 53 Variable BILEACIDUM Type Numeric Length 8 Format . Label Total serum bile acids (umol/L)

Variable Number 54 Variable BILEACIDUG Type Numeric Length 8 Format . Label Total serum bile acids (ug/dL)

Variable Number 55 Variable INR Type Numeric Length 8 Format . Label INR

Variable Number 56 Variable VITAMIND Type Numeric Length 8 Format . Label Vitamin D (ng/mL)

Variable Number 57 Variable ALKPHOS Type Numeric Length 8 Format . Label Alkaline phosphatase (U/L)

Variable Number 58 Variable GGT Type Numeric Length 8 Format . Label GGT (U/L)

Variable Number 59 Variable ALT Type Numeric Length 8 Format . Label ALT (U/L)

Variable Number 60 Variable AST Type Numeric Length 8 Format . Label AST (U/L)

Variable Number 61 Variable WBC Type Numeric Length 8 Format . Label WBC

Variable Number 62 Variable HGB Type Numeric Length 8 Format . Label Hemoglobin (g/dl)



Variable Number 63 Variable PLATE Type Numeric Length 8 Format . Label Platelets

Variable Number 64 Variable BONEFRACTUREANY Type Numeric Length 8 Format . Label any bone fracture

Variable Number 65 Variable BONEFRACTUREGT1 Type Numeric Length 8 Format . Label more than one bone fracture

Variable Number 66 Variable BONEFRACTURENUM Type Numeric Length 8 Format . Label Number of bone fractures

Variable Number 67 Variable DIVERSIONFLAG Type Numeric Length 8 Format . Label Biliary diversion prior to scan

Variable Number 68 Variable PHYSICALEXAMAGEYEARS Type Numeric Length 8 Format . Label Age (years) at the time of height/weight measurement

Variable Number 69 Variable PHYSICALEXAMAGEMONTHS Type Numeric Length 8 Format . Label Age (months) at the time of height/weight measurement

Variable Number 70 Variable BMI Type Numeric Length 8 Format . Label BMI closest to scan

Variable Number 71 Variable WEIGHTZSCORE Type Numeric Length 8 Format . Label Weight Z-score

Variable Number 72 Variable BMIZSCORE Type Numeric Length 8 Format . Label BMI Z-score

Variable Number 73 Variable HEIGHTZSCORE Type Numeric Length 8 Format . Label Height Z-Score

Variable Number 74 Variable SCANYEAR Type Numeric Length 8 Format . Label Year of DXA scan

Variable Number 75 Variable LEANBODYMASSKG Type Numeric Length 8 Format . Label Calculated lean body mass (kg)

Variable Number 76 Variable LEANBODYMASSPERCENT Type Numeric Length 8 Format . Label Calculated lean body mass (percent)

Variable Number 77 Variable DIVERSIONTOSCANYRS Type Numeric Length 8 Format . Label Time (years) from biliary diversion to DXA scan

Variable Number 78 Variable DIAGNOSISORDER Type Numeric Length 8 Format . Label Alternate ordering of diagnosis categories

Variable Number 79 Variable BMDTOTALZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body BMD Z-score

Variable Number 80 Variable BMDTOTALWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body BMD Z-score adjusted for weight, height

Variable Number 81 Variable BMDTOTALWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body BMD Z-score adjusted for weight, height, fat

Variable Number 82 Variable BMCTOTALZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body BMC Z-score

Variable Number 83 Variable BMCTOTALWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body BMC Z-score adjusted for weight, height

Variable Number 84 Variable BMCTOTALWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body BMC Z-score adjusted for weight, height, fat

Variable Number 85 Variable BMDSUBTOTALZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body minus head BMD Z-score

Variable Number 86 Variable BMDSUBTOTALWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body minus head BMD Z-score adjusted for weight, height

Variable Number 87 Variable BMDSUBTOTALWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body minus head BMD Z-score adjusted for weight, height, fat

Variable Number 88 Variable BMCSUBTOTALZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body minus head BMC Z-score

Variable Number 89 Variable BMCSUBTOTALWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body minus head BMC Z-score adjusted for weight, height

Variable Number 90 Variable BMCSUBTOTALWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Whole body minus head BMC Z-score adjusted for weight, height, fat

Variable Number 91 Variable BMDSPINEZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total spine BMD Z-score

Variable Number 92 Variable BMDSPINEWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total spine BMD Z-score adjusted for weight, height

Variable Number 93 Variable BMDSPINEWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total spine BMD Z-score adjusted for weight, height, fat

Variable Number 94 Variable BMCSPINEWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total spine BMC Z-score adjusted for weight, height

Variable Number 95 Variable BMCSPINEWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total spine BMC Z-score adjusted for weight, height, fat

Variable Number 96 Variable BMDNECKZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Hip neck BMD Z-score

Variable Number 97 Variable BMDNECKWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Hip neck BMD Z-score adjusted for weight, height

Variable Number 98 Variable BMDNECKWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Hip neck BMD Z-score adjusted for weight, height, fat

Variable Number 99 Variable BMCNECKWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Hip neck BMC Z-score adjusted for weight, height

Variable Number 100 Variable BMCNECKWTHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Hip neck BMC Z-score adjusted for weight, height, fat

Variable Number 101 Variable BMDHIPZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total hip BMD Z-score

Variable Number 102 Variable BMDHIPWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total hip BMD Z-score adjusted for weight, height

Variable Number 103 Variable BMDHIPWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total hip BMD Z-score adjusted for weight, height, fat

Variable Number 104 Variable BMCHIPWHTZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total hip BMC Z-score adjusted for weight, height

Variable Number 105 Variable BMCHIPWHTFATZSCORE\_ORIG Type Numeric Length 8 Format . Label Non-Winsorized Total hip BMC Z-score adjusted for weight, height, fat

Variable Number 106 Variable BMDTOTALZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDTotalZScore Winsorized?

Variable Number 107 Variable BMDTOTALWHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDTotalWtHtZScore Winsorized?

Variable Number 108 Variable BMDTOTALWHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDTotalWtHtFatZScore Winsorized?

Variable Number 109 Variable BMCTOTALZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCTotalZScore Winsorized?

Variable Number 110 Variable BMCTOTALWHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCTotalWtHtZScore Winsorized?

Variable Number 111 Variable BMCTOTALWHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCTotalWtHtFatZScore Winsorized?

Variable Number 112 Variable BMDSUBTOTALZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDSubTotalZScore Winsorized?

Variable Number 113 Variable BMDSUBTOTALWHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDSubTotalWtHtZScore Winsorized?

Variable Number 114 Variable BMDSUBTOTALWHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDSubTotalWtHtFatZScore Winsorized?

Variable Number 115 Variable BMCSUBTOTALZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCSubTotalZScore Winsorized?

Variable Number 116 Variable BMCSUBTOTALWHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCSubTotalWtHtZScore Winsorized?

Variable Number 117 Variable BMCSUBTOTALWTHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCSUBTotalWtHtFatZScore Winsorized?

Variable Number 118 Variable BMDSPINEZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDSpineZScore Winsorized?

Variable Number 119 Variable BMDSPINEWTHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDSpineWtHtZScore Winsorized?

Variable Number 120 Variable BMDSPINEWTHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDSpineWtHtFatZScore Winsorized?

Variable Number 121 Variable BMCSPINEWTHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCSpineWtHtZScore Winsorized?

Variable Number 122 Variable BMCSPINEWTHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCSpineWtHtFatZScore Winsorized?

Variable Number 123 Variable BMDNECKZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDNeckZScore Winsorized?

Variable Number 124 Variable BMDNECKWTHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDNeckWtHtZScore Winsorized?

Variable Number 125 Variable BMDNECKWTHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDNeckWtHtFatZScore Winsorized?

Variable Number 126 Variable BMCNECKWTHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCNeckWtHtZScore Winsorized?

Variable Number 127 Variable BMCNECKWTHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCNeckWtHtFatZScore Winsorized?

Variable Number 128 Variable BMDHIPZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDHipZScore Winsorized?

Variable Number 129 Variable BMDHIPWTHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDHipWtHtZScore Winsorized?

Variable Number 130 Variable BMDHIPWTHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMDHipWtHtFatZScore Winsorized?

Variable Number 131 Variable BMCHIPWTHTZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCHipWtHtZScore Winsorized?

Variable Number 132 Variable BMCHIPWTHTFATZSCORE\_WINFLAG Type Numeric Length 8 Format . Label Was BMCHipWtHtFatZScore Winsorized?

Variable Number 133 Variable TANNERSTAGEDEVELOP Type Numeric Length 8 Format . Label Tanner score - development

Variable Number 134 Variable TANNERSTAGEHAIR Type Numeric Length 8 Format . Label Tanner score - pubic hair

Variable Number 135 Variable SPLEENPLATE Type Numeric Length 8 Format . Label Spleen > 2cm and Platelet count <150

Variable Number 136 Variable BMILTNEG2 Type Numeric Length 8 Format . Label BMI Z-score < -2

Variable Number 137 Variable BONERISKYEARS Type Numeric Length 8 Format . Label Time (years) at risk for bone fractures

Variable Number 138 Variable CENTRALBILEACID Type Numeric Length 8 Format . Label Total serum bile acids (umol/L)

Variable Number 139 Variable GENETICDGN Type Numeric Length 8 Format . Label Genetic diagnosis

### Data Set Name: [manuscript\\_data\\_03oct19.sas7bdat](#)

Variable Number 1 Variable ChiLDReN\_StudyID Type Character Length 50 Format \$50. Label ChiLDReN\_StudyID

Variable Number 2 Variable sex Type Numeric Length 8 Format SEXF. Label Sex

Variable Number 3 Variable Ethnicity Type Numeric Length 8 Format ETHNF. Label Ethnicity

Variable Number 4 Variable dx Type Numeric Length 8 Format DXF. Label Diagnosis

Variable Number 5 Variable age\_consent\_y Type Numeric Length 8 Format NONE Label Age at consent (years)

Variable Number 6 Variable VCI Type Numeric Length 8 Format 11. Label Verbal Comprehension

Variable Number 7 Variable PRI Type Numeric Length 8 Format 11. Label Perceptual Reasoning

Variable Number 8 Variable WMI Type Numeric Length 8 Format 11. Label Working Memory

Variable Number 9 Variable PSI Type Numeric Length 8 Format 11. Label Processing Speed

Variable Number 10 Variable FSIQ Type Numeric Length 8 Format 11. Label Full Scale IQ

Variable Number 11 Variable VIQ Type Numeric Length 8 Format 11. Label Verbal IQ

Variable Number 12 Variable PIQ Type Numeric Length 8 Format 11. Label Performance IQ

Variable Number 13 Variable GLC Type Numeric Length 8 Format 11. Label General Language Component

Variable Number 14 Variable age\_test\_y Type Numeric Length 8 Format NONE Label Age at testing (years)

Variable Number 15 Variable race Type Numeric Length 8 Format RACEF. Label Race

Variable Number 16 Variable CEPH Type Numeric Length 8 Format CEPHF. Label CEPH

Variable Number 17 Variable college Type Numeric Length 8 Format COLLEGEF. Label Parental education

Variable Number 18 Variable xanthoma Type Numeric Length 8 Format PRESENTF. Label History of xanthoma(s)

Variable Number 19 Variable waz Type Numeric Length 8 Format NONE Label Weight-for-age z-score

Variable Number 20 Variable bmiz Type Numeric Length 8 Format NONE Label BMI z-score

Variable Number 21 Variable haz Type Numeric Length 8 Format NONE Label Height-for-age z-score

Variable Number 22 Variable pruritus Type Numeric Length 8 Format PRURITUSF. Label Pruritus

Variable Number 23 Variable INR Type Numeric Length 8 Format 8.1 Label INR

Variable Number 24 Variable AlbuminGdl Type Numeric Length 8 Format NONE Label Albumin (g/dl)

Variable Number 25 Variable BUNMgdl Type Numeric Length 8 Format NONE Label BUN (mg/dl)

Variable Number 26 Variable CreatinineMgdl Type Numeric Length 8 Format NONE Label Creatinine (mg/dl)

Variable Number 27 Variable HgbGdl Type Numeric Length 8 Format NONE Label Hemoglobin (g/dl)

Variable Number 28 Variable TotalBilirubinMgdl Type Numeric Length 8 Format NONE Label Total bilirubin (mg/dl)

Variable Number 29 Variable AlkPhosphUnitsL Type Numeric Length 8 Format BEST12. Label Alkaline phosphatase (U/L)

Variable Number 30 Variable ALTUnitsL Type Numeric Length 8 Format BEST12. Label ALT (U/L)

Variable Number 31 Variable ASTUnitsL Type Numeric Length 8 Format BEST12. Label AST (U/L)

Variable Number 32 Variable GGTPUnitsL Type Numeric Length 8 Format BEST12. Label GGTP (U/L)

Variable Number 33 Variable PlateletsCnt Type Numeric Length 8 Format BEST12. Label Platelet count

Variable Number 34 Variable APRI Type Numeric Length 8 Format NONE Label APRI

Variable Number 35 Variable FIB\_4 Type Numeric Length 8 Format NONE Label FIB-4

Variable Number 36 Variable cardiac\_cat Type Numeric Length 8 Format CARDIAC\_CATF. Label Cardiac disease severity

Variable Number 37 Variable JAG1 Type Numeric Length 8 Format YN. Label JAG1 mutation

Variable Number 38 Variable jag1combo Type Numeric Length 8 Format GENEF. Label JAG1 mutation

Variable Number 39 Variable ATP8B1combo Type Numeric Length 8 Format GENEF. Label ATP8B1 mutation

Variable Number 40 Variable ABCB11combo Type Numeric Length 8 Format GENEF. Label ABCB11 mutation

Variable Number 41 Variable ABCB4combo Type Numeric Length 8 Format GENE. Label ABCB4 mutation

Variable Number 42 Variable A1ATPhenoCombo Type Numeric Length 8 Format PHENOF. Label A1AT Phenotype

Variable Number 43 Variable IHxAnyRenalAbnormal Type Numeric Length 8 Format PRESENTF. Label Any renal abnormality

Variable Number 44 Variable time\_fu\_y Type Numeric Length 8 Format NONE Label Follow-up time (years)

Variable Number 45 Variable tested Type Numeric Length 8 Format YN. Label Tested

Variable Number 46 Variable test Type Numeric Length 8 Format TESTF. Label Test type

### Data Set Name: parent\_univariate.sas7bdat

Variable Number 1 Variable CTXSTDID Type Numeric Length 8 Format . Label De-Identified Subject ID

Variable Number 2 Variable PHYSICAL Type Numeric Length 8 Format . Label QOL Physical

Variable Number 3 Variable PSYCHOSOCIAL Type Numeric Length 8 Format . Label QOL psychosocial

Variable Number 4 Variable TOTAL Type Numeric Length 8 Format . Label QOL Total

Variable Number 5 Variable WAZ Type Numeric Length 8 Format . Label Weight z-scores

Variable Number 6 Variable HAZ Type Numeric Length 8 Format . Label Height z-scores

Variable Number 7 Variable INR\_N Type Numeric Length 8 Format . Label INR

Variable Number 8 Variable DIS\_GRP Type Character Length 25 Format . Label Disease Group - ALGS, BAD, PFIC, a1-AT

Variable Number 9 Variable GRP Type Numeric Length 8 Format . Label Eligibility group

Variable Number 10 Variable AGE\_AT\_TEST Type Numeric Length 8 Format . Label Age at QOL survey (years)

Variable Number 11 Variable CARDIAC\_DEFECT Type Character Length 1 Format . Label Cardiac Defect - 1=No Defect, 2=Other Defect, 3=PPS

### Data Set Name: pedsq\_child\_dis.sas7bdat

Variable Number 1 Variable DIS\_GRP Type Character Length 25 Format . Label Disease Group - ALGS, BAD, PFIC, a1-AT

Variable Number 2 Variable CTXSTDID Type Numeric Length 8 Format . Label De-Identified Subject ID

Variable Number 3 Variable GRP Type Numeric Length 8 Format . Label Eligibility group

Variable Number 4 Variable FROM Type Character Length 20 Format . Label QOL form source

Variable Number 5 Variable PHYSICAL Type Numeric Length 8 Format . Label QOL Physical  
Variable Number 6 Variable EMOTION Type Numeric Length 8 Format . Label QOL emotion  
Variable Number 7 Variable SOCIAL Type Numeric Length 8 Format . Label QOL social  
Variable Number 8 Variable SCHOOL Type Numeric Length 8 Format . Label QOL school  
Variable Number 9 Variable PSYCHOSOCIAL Type Numeric Length 8 Format . Label QOL psychosocial  
Variable Number 10 Variable TOTAL Type Numeric Length 8 Format . Label QOL Total

Data Set Name: pedsq1\_parent\_dis.sas7bdat

Variable Number 1 Variable DIS\_GRP Type Character Length 25 Format . Label Disease Group - ALGS, BAD, PFIC, a1-AT

Variable Number 2 Variable CTXSTDID Type Numeric Length 8 Format . Label De-Identified Subject ID

Variable Number 3 Variable GRP Type Numeric Length 8 Format . Label Eligibility group

Variable Number 4 Variable FROM Type Character Length 21 Format . Label QOL form source

Variable Number 5 Variable PHYSICAL Type Numeric Length 8 Format . Label QOL Physical

Variable Number 6 Variable EMOTION Type Numeric Length 8 Format . Label QOL emotion

Variable Number 7 Variable SOCIAL Type Numeric Length 8 Format . Label QOL social

Variable Number 8 Variable SCHOOL Type Numeric Length 8 Format . Label QOL school

Variable Number 9 Variable PSYCHOSOCIAL Type Numeric Length 8 Format . Label QOL psychosocial

Variable Number 10 Variable TOTAL Type Numeric Length 8 Format . Label QOL Total

### Data Set Name: pfic.sas7bdat

Variable Number 1 Variable bmi Type Numeric Length 8 Format NONE Label BMI at baseline exam

Variable Number 2 Variable WeightZScore Type Numeric Length 8 Format NONE Label weight-for-age Z at baseline exam

Variable Number 3 Variable BMIZScore Type Numeric Length 8 Format NONE Label BMI-for-age Z at baeline exam

Variable Number 4 Variable HeightZScore Type Numeric Length 8 Format NONE Label height-for-age Z at baeline exam

Variable Number 5 Variable Gender Type Numeric Length 8 Format SEXFMT. Label Sex

Variable Number 6 Variable Ethnicity Type Numeric Length 8 Format ETHNICITYFMT. Label Ethnicity

Variable Number 7 Variable SubjectID Type Character Length 8 Format NONE Label LOGIC subject ID



Variable Number 8 Variable PrimaryDgn Type Numeric Length 8 Format PRIMARYDGNFMT. Label Primary diagnosis

Variable Number 9 Variable LogicGroup Type Numeric Length 8 Format LOGICGROUPEFMT. Label LOGIC group

Variable Number 10 Variable Race Type Numeric Length 8 Format RACEFMT. Label Race

Variable Number 11 Variable AgeAtConsent Type Numeric Length 8 Format NONE Label Age at consent (years)

Variable Number 12 Variable DrainAny Type Numeric Length 8 Format YESNOMISSFMT. Label Any drainage procedure performed

Variable Number 13 Variable DrainSelect Type Numeric Length 8 Format YESNOMISSFMT. Label Subject had Choledochojejunostomy, Biliary diversion or Ileal exclusion any time during study

Variable Number 14 Variable SpleenSize Type Numeric Length 8 Format NONE Label Spleen size (cm) below costal margin (set to 0 if not palpable)

Variable Number 15 Variable HeightCm Type Numeric Length 8 Format NONE Label Height (cm) at baseline exam

Variable Number 16 Variable WeightKg Type Numeric Length 8 Format NONE Label Weight (kg) at baseline exam

Variable Number 17 Variable Pruritus Type Numeric Length 8 Format PRURITUSFMT. Label Pruritus at baseline physical exam

Variable Number 18 Variable Xanthoma Type Numeric Length 8 Format XANTHOMAFMT. Label Xanthoma at baseline exam

Variable Number 19 Variable INR Type Numeric Length 8 Format NONE Label INR at baseline exam

Variable Number 20 Variable AlbuminGdl Type Numeric Length 8 Format NONE Label Albumin (g/dL) at baseline exam

Variable Number 21 Variable SerumBileAcidsUmolL Type Numeric Length 8 Format NONE Label Total serum bile acids (umol/L) at baseline exam

Variable Number 22 Variable CalciumMgdl Type Numeric Length 8 Format NONE Label Calcium (mg/dL) at baseline exam

Variable Number 23 Variable CholesterolMgdl Type Numeric Length 8 Format NONE Label Total cholesterol (mg/dL) at baseline exam

Variable Number 24 Variable CreatinineMgdl Type Numeric Length 8 Format NONE Label Creatinine (mg/dL) at baseline exam

Variable Number 25 Variable GlucoseMgdl Type Numeric Length 8 Format NONE Label Glucose (mg/dL) at baseline exam

Variable Number 26 Variable HgbGdl Type Numeric Length 8 Format NONE Label Hemoglobin (g/dL) at baseline exam

Variable Number 27 Variable PhosphorusMgdI Type Numeric Length 8 Format NONE Label Phosphorus (mg/dL) at baseline exam

Variable Number 28 Variable RBPMgdI Type Numeric Length 8 Format NONE Label Retinol binding protein (mg/dL) at baseline exam

Variable Number 29 Variable SerumLipidsMgdI Type Numeric Length 8 Format NONE Label Total serum lipids (mg/dL) at baseline exam

Variable Number 30 Variable TotalProteinGdl Type Numeric Length 8 Format NONE Label Total protein (g/dL) at baseline exam

Variable Number 31 Variable VitaminAUgdI Type Numeric Length 8 Format NONE Label Vitamin A (ug/dL) at baseline exam

Variable Number 32 Variable VitaminD125OH2Pgml Type Numeric Length 8 Format NONE Label 1,25 OH2 Vitamin D (pg/mL) at baseline exam

Variable Number 33 Variable VitaminD250HNgml Type Numeric Length 8 Format NONE Label Vitamin D (D25OH) (ng/mL) at baseline exam

Variable Number 34 Variable VitaminEUgdI Type Numeric Length 8 Format NONE Label Vitamin E (ug/mL) at baseline exam

Variable Number 35 Variable AlkPhosphUnitsL Type Numeric Length 8 Format NONE Label Alkaline phosphate (U/L) at baseline exam

Variable Number 36 Variable ALTUnitsL Type Numeric Length 8 Format NONE Label ALT (U/L) at baseline exam

Variable Number 37 Variable ASTUnitsL Type Numeric Length 8 Format NONE Label AST (U/L) at baseline exam

Variable Number 38 Variable CIMmolL Type Numeric Length 8 Format NONE Label Chloride (mmol/L) at baseline exam

Variable Number 39 Variable CO2MmolL Type Numeric Length 8 Format NONE Label Bicarbonate (mmol/L) at baseline exam

Variable Number 40 Variable GGTPUnitsL Type Numeric Length 8 Format NONE Label GGTP (U/L) at baseline exam

Variable Number 41 Variable KMmolL Type Numeric Length 8 Format NONE Label Potassium (mmol/L) at baseline exam

Variable Number 42 Variable NaMmolL Type Numeric Length 8 Format NONE Label Sodium (mmol/L) at baseline exam

Variable Number 43 Variable PlateletsCnt Type Numeric Length 8 Format NONE Label Platelets (10/mm) at baseline exam

Variable Number 44 Variable WBCcnt Type Numeric Length 8 Format NONE Label White blood cells (10/mm) at baseline exam

Variable Number 45 Variable BiliTotalMgdl Type Numeric Length 8 Format NONE Label Total bilirubin (mg/dL) at baseline exam

Variable Number 46 Variable RBC Type Numeric Length 8 Format NONE Label Red blood cells (10/mm) at baseline exam

Variable Number 47 Variable AgeAtExam Type Numeric Length 8 Format NONE Label Age at exam (years) at baseline exam

Variable Number 48 Variable TimeFromSymptomOnset Type Numeric Length 8 Format NONE Label Time (years) from onset of first disease symptoms to baseline exam

Variable Number 49 Variable TDDrainSelect Type Numeric Length 8 Format YESNOMISSFMT. Label Subject had Choledochojejunostomy, Biliary diversion or Ileal exclusion on or before exam

Variable Number 50 Variable AgeAtTxp Type Numeric Length 8 Format NONE Label Age (years) at liver transplant

Variable Number 51 Variable AgeAtBaseline Type Numeric Length 8 Format NONE Label Age (years) at baseline LOGIC visit

Variable Number 52 Variable AgeAtFirstSymptomYrs Type Numeric Length 8 Format NONE Label Age (years) at onset of first liver disease symptoms

Variable Number 53 Variable AgeAtFirstDgnYrs Type Numeric Length 8 Format NONE Label Age (years) at initial diagnosis of liver disease (from initial history CRF)

Variable Number 54 Variable TxpTimeYrs Type Numeric Length 8 Format NONE Label Time (years) from LOGIC consent to liver transplant (groups 1 and 2 only)

Variable Number 55 Variable GeneticDgn Type Numeric Length 8 Format GENETICDGNFMT. Label Genetic diagnosis

Variable Number 56 Variable nMutations Type Numeric Length 8 Format NONE Label nMutations

Variable Number 57 Variable MutationCode Type Character Length 7 Format \$7. Label MutationCode

Variable Number 58 Variable GeneticGroup Type Numeric Length 8 Format GENETICGROUPEFMT. Label GeneticGroup

Variable Number 59 Variable dgnSpecifyOther Type Character Length 200 Format \$200. Label Other, specify diagnosis field

Variable Number 60 Variable DaysToSelectDrain Type Numeric Length 8 Format NONE Label Number of days from baseline to drainage procedure

Variable Number 61 Variable DaysToTransplant Type Numeric Length 8 Format NONE Label Number of days from baseline to transplant

Variable Number 62 Variable AgeAtConsentMo Type Numeric Length 8 Format NONE Label Age (months) at enrollment

Variable Number 63 Variable AgeAtFirstDgnMo Type Numeric Length 8 Format NONE Label Age (months) at diagnosis

Variable Number 64 Variable AgeAtFirstSymptomMo Type Numeric Length 8 Format NONE Label Age (months) at onset of first liver disease symptoms

Variable Number 65 Variable BLUrso Type Numeric Length 8 Format YESNOMISSFMT. Label Urso at baseline

Variable Number 66 Variable PruritusAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of pruritus on or before baseline exam

Variable Number 67 Variable JaundiceAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of jaundice on or before baseline exam

Variable Number 68 Variable DiarrheaAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of diarrhea on or before baseline exam

Variable Number 69 Variable Failure2ThriveAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of failure to thrive on or before baseline exam

Variable Number 70 Variable BleedingAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of bleeding or bruising on or before baseline exam

Variable Number 71 Variable DiureticsAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label Ascites at baseline based on diuretic use

Variable Number 72 Variable GIBleedAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of GI bleeding on or before baseline exam

Variable Number 73 Variable BoneFractureAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of bone fracture on or before baseline exam

Variable Number 74 Variable RicketsAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of rickets on or before baseline exam

Variable Number 75 Variable GallstonesAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of gallstones on or before baseline exam

Variable Number 76 Variable HearingProbAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label History of hearing problems on or before baseline exam

Variable Number 77 Variable PreBLAndTxpSelectDrain Type Numeric Length 8 Format YESNOMISSFMT. Label Drainage procedure performed prior to baseline (or transplant for post-transplant participants)

Variable Number 78 Variable PruritusRxAtBL Type Numeric Length 8 Format YESNOMISSFMT. Label Pruritus Rx at baseline

Variable Number 79 Variable PreBLGt1YrSelectDrain Type Numeric Length 8 Format YESNOMISSFMT. Label Drain placed at least one year before baseline

Variable Number 80 Variable PerfectDrain Type Numeric Length 8 Format YESNOMISSFMT. Label Bili le 1, no pruritus meds, minimal or no pruritus, platelets at least 150, spleen lt 2cm

Variable Number 81 Variable SuccessfulDrain Type Numeric Length 8 Format YESNOMISSFMT. Label Bili le 1, no pruritus meds, minimal or no pruritus, platelets at least 150, spleen lt 2cm

Variable Number 82 Variable One Type Numeric Length 8 Format NONE Label Variable with value of 1

Variable Number 83 Variable MutationCodeNum Type Numeric Length 8 Format MUTATIONCODENUMFMT. Label Detailed mutation group

Variable Number 84 Variable FirstBaseline Type Numeric Length 8 Format MMDDYY10. Label Earliest enrollment date for participants in the study

Variable Number 85 Variable LastBaseline Type Numeric Length 8 Format MMDDYY10. Label Latest enrollment date for participants in the study

### Data Set Name: [pficgenetics.sas7bdat](#)

Variable Number 1 Variable SubjectID Type Character Length 8 Format NONE Label LOGIC subject ID

Variable Number 2 Variable GeneticGroup Type Numeric Length 8 Format GENETICGROUPEFMT. Label GeneticGroup

Variable Number 3 Variable LogicGroup Type Numeric Length 8 Format LOGICGROUPEFMT. Label LOGIC group

Variable Number 4 Variable Gene Type Numeric Length 8 Format GENEFMT. Label Gene

Variable Number 5 Variable MutationYN Type Numeric Length 8 Format YNUFMT. Label Mutation status

Variable Number 6 Variable TestDate Type Numeric Length 8 Format MMDDYY8. Label B-1 DNA Test Date:

Variable Number 7 Variable cDNAChange Type Numeric Length 8 Format YESNOMISSFMT. Label B-5: Mutation Detected : 1: cDNA change, specify:

Variable Number 8 Variable Mutation1\_Other Type Character Length 200 Format \$50. Label B-5: Mutation Detected : 1: cDNA change, specify: [text]

Variable Number 9 Variable ProteinChange Type Numeric Length 8 Format YESNOMISSFMT. Label B-5: Mutation Detected : 2: Protein change, specify:

Variable Number 10 Variable Mutation2\_Other Type Character Length 200 Format \$50. Label B-5: Mutation Detected : 2: Protein change, specify: [text]

Variable Number 11 Variable MutChangeType Type Numeric Length 8 Format MUTCHANGETYPEFMT.  
Label B:6 Change Type:

Variable Number 12 Variable DiseaseCause Type Numeric Length 8 Format DISEASECAUSEFMT. Label B:7  
Disease Causing: