

ALGS Neuro Codebook

Variable Name	Variable Label	Variable Format or Range
ChiLDReN_StudyID	De-Identified Subject ID	
sex	Sex	1=Male; 2=Female
Ethnicity	Ethnicity	1=Hispanic; 2=Not Hispanic
dx	Diagnosis	1=ALGS; 2=PFIC; 3=A1AT
age_consent_y	Age at consent (years)	0.1-17.0
VCI	Verbal Comprehension	47-140
PRI	Perceptual Reasoning	45-149
WMI	Working Memory	54-126
PSI	Processing Speed	50-123
FSIQ	Full Scale IQ	41-132
VIQ	Verbal IQ	70-139
PIQ	Performance IQ	63-129
GLC	General Language Component	70-133
age_test_y	Age at testing (years)	2.7-16.9
race	Race	1=White; 2=Black; 3=Other
CEPH	CEPH	0=aCEPH; 1=pCEPH; 2=dCEPH
college	Parental education	0=High school equivalent or less; 1=Some college/trade school or more
xanthoma	History of xanthoma(s)	0=Absent; 1=Present
waz	Weight-for-age z-score	-4.4-3.7
bmiz	BMI z-score	-5.3-4.2
haz	Height-for-age z-score	-4.7-2.5
pruritus	Pruritus	0=None/Mild; 1=Active/Bleeding
INR	INR	0.82-2.7
AlbuminGdl	Albumin (g/dl)	2.1-5.4
BUNMgdl	BUN (mg/dl)	6-62
CreatinineMgdl	Creatinine (mg/dl)	0.2-13
HgbGdl	Hemoglobin (g/dl)	8.5-19.5
TotalBilirubinMgdl	Total bilirubin (mg/dl)	0-23
AlkPhosphUnitsL	Alkaline phosphatase (U/L)	92-2540
ALTUnitsL	ALT (U/L)	2.5-552
ASTUnitsL	AST (U/L)	23-485
GGTPUnitsL	GGTP (U/L)	8-1585

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Variable Name	Variable Label	Variable Format or Range
PlateletsCnt	Platelet count	30-721
APRI	APRI	0.2-17.5
FIB_4	FIB-4	0.1-5.3
cardiac_cat	Cardiac disease severity	0=Mild/None; 1=Severe
JAG1	JAG1 mutation	0=No; 1=Yes
jag1combo	JAG1 mutation	-1=Testing not done; 0=No known mutations; 1=Total gene deletion; 2=Protein truncating; 3=Missense; 4=Splicing; 5=Synonymous; 6=Multiple mutation types
ATP8B1combo	ATP8B1 mutation	-1=Testing not done; 0=No known mutations; 1=Total gene deletion; 2=Protein truncating; 3=Missense; 4=Splicing; 5=Synonymous; 6=Multiple mutation types
ABCB11combo	ABCB11 mutation	-1=Testing not done; 0=No known mutations; 1=Total gene deletion; 2=Protein truncating; 3=Missense; 4=Splicing; 5=Synonymous; 6=Multiple mutation types
ABCB4combo	ABCB4 mutation	-1=Testing not done; 0=No known mutations; 1=Total gene deletion; 2=Protein truncating; 3=Missense; 4=Splicing; 5=Synonymous; 6=Multiple mutation types
A1ATPhenoCombo	A1AT Phenotype	1=ZZ; 2=SZ; 3=Unknown
IHxAnyRenalAbnormal	Any renal abnormality	0=Absent; 1=Present
time_fu_y	Follow-up time (years)	0.0-10.3
tested	Tested	0=No; 1=Yes
test	Test type	0=WPPSI-III (age 3 to 5); 1=WISC-IV (age 6 to 16)