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## **Online Supplementary Material**

Glycogen Storage Disease Type Ia: A Retrospective Claims Analysis of Complications, Resource Utilization, and Cost of Care. *JHEOR*. 2025;12(1):13-21. doi:10.36469/jheor.2025.125886

### Table S1: ICD-10-CM Codes Used to Identify Complications in Patients With GSDIa

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Figure S1: Complications in Children With GSDIa vs Comparators

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Figure S4: Frequency of Hospitalizations by Patient Age in Patients With GSDIa (A) and in Comparators (B)

This supplementary material has been provided by the authors to give readers additional information about their work.



Complication	ICD-10-D Codes					
GSDIa	ICD-10-D-E7401, excluding patients with inflammatory bowel disease					
Digestive System						
Abdominal Pain	ICD-10-D-R1010, ICD-10-D-R1011, ICD-10-D-R1012, ICD-10-D- R1013, ICD-10-D-R1030, ICD-10-D-R1031, ICD-10-D-R1032, ICD-10- D-R1033, ICD-10-D-R1084, ICD-10-D-R109 ICD-10-D-R197					
Diarrhea						
Gallstones	ICD-10-D-K8000, ICD-10-D-K8001, ICD-10-D-K8010, ICD-10-D-K8011, ICD-10-D-K8012, ICD-10-D-K8013, ICD-10-D-K8018, ICD-10-D-K8019, ICD-10-D-K8020, ICD-10-D-K8021, ICD-10-D-K8030, ICD-10-D-K8031, ICD-10-D-K8032, ICD-10-D-K8033, ICD-10-D-K8034, ICD-10-D-K8035, ICD-10-D-K8037, ICD-10-D-K8040, ICD-10-D-K8041, ICD-10-D-K8042, ICD-10-D-K8043, ICD-10-D-K8044, ICD-10-D-K8045, ICD-10-D-K8046, ICD-10-D-K8047, ICD-10-D-K8050, ICD-10-D-K8051, ICD-10-D-K8060, ICD-10-D-K8061, ICD-10-D-K8062, ICD-10-D-K8063, ICD-10-D-K8064, ICD-10-D-K8065, ICD-10-D-K8066, ICD-10-D-K8067, ICD-10-D-K8070, ICD-10-D-K8071, ICD-10-D-K8080, ICD-10-D-K8080, ICD-10-D-K8080, ICD-10-D-K8080, ICD-10-D-K8081					
Gastroesophageal reflux disease	ICD-10-D-K210, ICD-10-D-K219					
Intestinal gas	ICD-10-D-R140, ICD-10-D-R141, ICD-10-D-R142, ICD-10-D-R143					
Nausea and/or Vomiting	ICD-10-D-R110, ICD-10-D-R1110, ICD-10-D-R1111, ICD-10-D- R1112, ICD-10-D-R112					
Pancreatitis	ICD-10-D-K850, ICD-10-D-K8500, ICD-10-D-K8501, ICD-10-D-K8502, ICD-10-D-K851, ICD-10-D-K8510, ICD-10-D-K8511, ICD-10-D-K8512, ICD-10-D-K852, ICD-10-D-K8520, ICD-10-D-K8521, ICD-10-D-K8522, ICD-10-D-K853, ICD-10-D-K8530, ICD-10-D-K8531, ICD-10-D-K8532, ICD-10-D-K858, ICD-10-D-K8580, ICD-10-D-K8581, ICD-10-D-K8582, ICD-10-D-K859, ICD-10-D-K8590, ICD-10-D-K8591, ICD-10-D-K8592, ICD-10-D-K860, ICD-10-D-K861					
Nutritional						
Anorexia	ICD-10-D-R630					
Gastrostomy	ICD-10-D-K9420, ICD-10-D-K9421, ICD-10-D-K9422, ICD-10-D- K9423, ICD-10-D-K9429, ICD-10-D-Z431, ICD-10-D-Z931					
Iron deficiency Nutritional deficiency	ICD-10-D-D500, ICD-10-D-D508, ICD-10-D-D509, ICD-10-D-E611 ICD-10-D-E559, ICD-10-D-D500, ICD-10-D-D508, ICD-10-D-D509, ICD-10-D-E611, ICD-10-D-D530, ICD-10-D-E40, ICD-10-D-E41, ICD- 10-D-E42, ICD-10-D-E43, ICD-10-D-E440, ICD-10-D-E441, ICD-10-D- E45, ICD-10-D-E46, ICD-10-D-E640, ICD-10-D-E500, ICD-10-D- E501, ICD-10-D-E502, ICD-10-D-E505, ICD-10-D-E506, ICD-10-D- E507, ICD-10-D-E508, ICD-10-D-E509, ICD-10-D-E5111, ICD-10-D- E5112, ICD-10-D-E512, ICD-10-D-E518, ICD-10-D-E519, ICD-10-D- E52, ICD-10-D-E530, ICD-10-D-E531, ICD-10-D-E538, ICD-10-D- E539, ICD-10-D-E54, ICD-10-D-E560, ICD-10-D-E5661, ICD-10-D- E568, ICD-10-D-E569, ICD-10-D-E58, ICD-10-D-E59, ICD-10-D-E60, ICD-10-D-E630, ICD-10-D-E631, ICD-10-D-E638, ICD-10-D-E639, ICD-10-D-E641, ICD-10-D-E642, ICD-10-D-E648, ICD-10-D-E649, ICD-10-D-511, ICD-10-D-513, ICD-10-D-518, ICD-10-D-519, ICD-10-D-520, ICD-10-D-534, ICD-10-D-538, ICD-10-D-539, ICD-10-D-554, ICD-10-D-553, ICD-10-D-558, ICD-10-D-559, ICD-10-D-559, ICD-10-D-554, ICD-10-D-553, ICD-10-D-558, ICD-10-D-559, ICD-10-D-5528, ICD-10-D-5529, ICD-10-D-5529, ICD-10-D-5529, ICD-10-D-559, IC					
Obesity	ICD-10-D-E6601, ICD-10-D-E6609, ICD-10-D-E662, ICD-10-D-E668, ICD-10-D-E669, ICD-10-D-Z6830, ICD-10-D-Z6831, ICD-10-D-Z6832,					

 Table S1. ICD-10-CM Codes Used to Identify Complications in Patients With GSDIa

Complication	ICD-10-D Codes
	ICD-10-D-Z6833, ICD-10-D-Z6834, ICD-10-D-Z6835, ICD-10-D-Z6836, ICD-10-D-Z6837, ICD-10-D-Z6838, ICD-10-D-Z6849, ICD-10-D-Z6841, ICD-10-D-Z6842, ICD-10-D-Z6843, ICD-10-D-Z6844, ICD-10-D-Z6845, ICD-10-D-Z6854
Other nutritional deficiency	ICD-10-D-E500, ICD-10-D-E501, ICD-10-D-E502, ICD-10-D-E505, ICD-10-D-E506, ICD-10-D-E507, ICD-10-D-E508, ICD-10-D-E509, ICD-10-D-E5111, ICD-10-D-E5112, ICD-10-D-E512, ICD-10-D-E518, ICD-10-D-E519, ICD-10-D-E52, ICD-10-D-E530, ICD-10-D-E531, ICD-10-D-E538, ICD-10-D-E539, ICD-10-D-E54, ICD-10-D-E560, ICD-10-D-E561, ICD-10-D-E568, ICD-10-D-E569, ICD-10-D-E58, ICD-10-D-E59, ICD-10-D-E60, ICD-10-D-E630, ICD-10-D-E631, ICD- 10-D-E638, ICD-10-D-E639, ICD-10-D-E641, ICD-10-D-E642, ICD-10- D-E648, ICD-10-D-E649, ICD-10-D-D511, ICD-10-D-D513, ICD-10-D- D518, ICD-10-D-D519, ICD-10-D-D520, ICD-10-D-D528, ICD-10-D- D529
Poor growth Protein-calorie malnutrition	ICD-10-D-R6251, ICD-10-D-R627, ICD-10-D-R620, ICD-10-D-R6250, ICD-10-D-R6252, ICD-10-D-R6259 ICD-10-D-D530, ICD-10-D-E40, ICD-10-D-E41, ICD-10-D-E42, ICD- 10-D-E43, ICD-10-D-E440, ICD-10-D-E441, ICD-10-D-E45, ICD-10-D- E46, ICD-10-D-E640
Vitamin D deficiency	ICD-10-D-E559
Hematologic	
Anemia Anemia due to enzyme disorders Iron deficiency anemia Acidosis	ICD-10-D-D500, ICD-10-D-D508, ICD-10-D-D509, ICD-10-D-D510, ICD-10-D-D511, ICD-10-D-D513, ICD-10-D-D518, ICD-10-D-D519, ICD-10-D-D520, ICD-10-D-D528, ICD-10-D-D529, ICD-10-D-D530, ICD-10-D-D531, ICD-10-D-D532, ICD-10-D-D538, ICD-10-D-D539, ICD-10-D-D630, ICD-10-D-D631, ICD-10-D-D638, ICD-10-D-D641, ICD-10-D-D643, ICD-10-D-D6481, ICD-10-D-D6489, ICD-10-D-D649 ICD-10-D-D550, ICD-10-D-D551, ICD-10-D-D552, ICD-10-D-D553, ICD-10-D-D558, ICD-10-D-D559 ICD-10-D-D500, ICD-10-D-D508, ICD-10-D-D509 ICD-10-D-E872
Cardiovascular/cardiopulmonary	
Atherosclerotic heart disease Hypertension	ICD-10-D-I2510, ICD-10-D-I25110, ICD-10-D-I25111, ICD-10-D- I25118, ICD-10-D-I25119 ICD-10-D-I10, ICD-10-D-I110, ICD-10-D-I119, ICD-10-D-I120, ICD-10- D-I129, ICD-10-D-I130, ICD-10-D-I131, ICD-10-D-I1310, ICD-10-D- I1311, ICD-10-D-I132, ICD-10-D-I150, ICD-10-D-I151, ICD-10-D-I152,
Pulmonary hypertension	ICD-10-D-I158, ICD-10-D-I159 ICD-10-D-I270, ICD-10-D-I272, ICD-10-D-I2720, ICD-10-D-I2721, ICD-10-D-I2722, ICD-10-D-I2729
Hepatic	
Hypoglycemia Hepatomegaly	ICD-10-D-E08641, ICD-10-D-E08649, ICD-10-D-E10641, ICD-10-D-E10649, ICD-10-D-E11641, ICD-10-D-E11649, ICD-10-D-E13641, ICD-10-D-E13649, ICD-10-D-E15, ICD-10-D-E160, ICD-10-D-E161, ICD-10-D-E162, ICD-10-D-P704 ICD-10-D-R160, ICD-10-D-R162
Hyperlipidemia	ICD-10-D-E780, ICD-10-D-E7800, ICD-10-D-E781, ICD-10-D-E782, ICD-10-D-E784, ICD-10-D-E785
Musculoskeletal	

Complication	ICD-10-D Codes
Fatigue	ICD-10-D-R530, ICD-10-D-R531, ICD-10-D-R538, ICD-10-D-R5381,
	ICD-10-D-R5382, ICD-10-D-R5383
Gout	ICD-10-D-M1000, ICD-10-D-M10011, ICD-10-D-M10012, ICD-10-D-
	M10021, ICD-10-D-M10031, ICD-10-D-M10032, ICD-10-D-M10039,
	ICD-10-D-M10041, ICD-10-D-M10042, ICD-10-D-M10061, ICD-10-D-
	M10062, ICD-10-D-M10069, ICD-10-D-M10071, ICD-10-D-M10072,
	ICD-10-D-M10079, ICD-10-D-M1009, ICD-10-D-M1030, ICD-10-D-
	M10332, ICD-10-D-M10361, ICD-10-D-M10362, ICD-10-D-M10371,
	ICD-10-D-M10372, ICD-10-D-M10379, ICD-10-D-M1039, ICD-10-D-
	M1040, ICD-10-D-M10461, ICD-10-D-M10462, ICD-10-D-M10471,
	ICD-10-D-M10472, ICD-10-D-M10479, ICD-10-D-M1049, ICD-10-D-
	M109, ICD-10-D-M1A00X0, ICD-10-D-M1A00X1, ICD-10-D-M1A0410,
	ICD-10-D-M1A0411, ICD-10-D-M1A0420, ICD-10-D-M1A0610, ICD-
	10-D-M1A0611, ICD-10-D-M1A0620, ICD-10-D-M1A0621, ICD-10-D-
	M1A0710, ICD-10-D-M1A0711, ICD-10-D-M1A0720, ICD-10-D-
	M1A0721, ICD-10-D-M1A0790, ICD-10-D-M1A09X0, ICD-10-D-
	M1A09X1, ICD-10-D-M1A30X0, ICD-10-D-M1A30X1, ICD-10-D-
	M1A3120, ICD-10-D-M1A3191, ICD-10-D-M1A3690, ICD-10-D-
	M1A3710, ICD-10-D-M1A3721, ICD-10-D-M1A38X1, ICD-10-D-
	M1A39X0, ICD-10-D-M1A39X1, ICD-10-D-M1A40X0, ICD-10-D-
	M1A4291, ICD-10-D-M1A4710, ICD-10-D-M1A4711, ICD-10-D-
	M1A4790, ICD-10-D-M1A9XX0, ICD-10-D-M1A9XX1
Osteoarthritis	ICD-10-D-M160, ICD-10-D-M1610, ICD-10-D-M1611, ICD-10-D-
	M1612, ICD-10-D-M162, ICD-10-D-M1630, ICD-10-D-M1631, ICD-10-
	D-M1632, ICD-10-D-M166, ICD-10-D-M167, ICD-10-D-M169, ICD-10-
	D-M170, ICD-10-D-M1710, ICD-10-D-M1711, ICD-10-D-M1712, ICD-
	10-D-M174, ICD-10-D-M175, ICD-10-D-M179, ICD-10-D-M180, ICD-
	10-D-M1810, ICD-10-D-M1811, ICD-10-D-M1812, ICD-10-D-M184,
	ICD-10-D-M1851, ICD-10-D-M1852, ICD-10-D-M189, ICD-10-D-
	M19011, ICD-10-D-M19012, ICD-10-D-M19019, ICD-10-D-M19021,
	ICD-10-D-M19022, ICD-10-D-M19029, ICD-10-D-M19031, ICD-10-D-
	M19032, ICD-10-D-M19039, ICD-10-D-M19041, ICD-10-D-M19042,
	ICD-10-D-M19049, ICD-10-D-M19071, ICD-10-D-M19072, ICD-10-D-
	M19079, ICD-10-D-M19211, ICD-10-D-M19212, ICD-10-D-M19219,
	ICD-10-D-M19221, ICD-10-D-M19222, ICD-10-D-M19231, ICD-10-D-
	M19241, ICD-10-D-M19242, ICD-10-D-M19249, ICD-10-D-M19271,
	ICD-10-D-M19272, ICD-10-D-M1990, ICD-10-D-M1991, ICD-10-D-
	M1993
Osteoporosis	ICD-10-D-M8000XA, ICD-10-D-M8000XD, ICD-10-D-M8000XG, ICD-
·	10-D-M8000XP, ICD-10-D-M8000XS, ICD-10-D-M80019D, ICD-10-D-
	M80021K, ICD-10-D-M80021S, ICD-10-D-M80022A, ICD-10-D-
	M80022G, ICD-10-D-M80022S, ICD-10-D-M80031A, ICD-10-D-
	M80051A, ICD-10-D-M80051D, ICD-10-D-M80051K, ICD-10-D-
	M80051S, ICD-10-D-M80052A, ICD-10-D-M80052D, ICD-10-D-
	M80059A, ICD-10-D-M80059S, ICD-10-D-M80061D, ICD-10-D-
	M80062A, ICD-10-D-M80071D, ICD-10-D-M80072A, ICD-10-D-
	M8008XA, ICD-10-D-M8008XD, ICD-10-D-M8008XG, ICD-10-D-
	M8008XS, ICD-10-D-M8080XA, ICD-10-D-M8080XD, ICD-10-D-
	M8080XG, ICD-10-D-M8080XK, ICD-10-D-M8080XS, ICD-10-D-
	M80811A, ICD-10-D-M80811D, ICD-10-D-M80812A, ICD-10-D-
	M80821A, ICD-10-D-M80821K, ICD-10-D-M80822A, ICD-10-D-
	M80822D, ICD-10-D-M80822K, ICD-10-D-M80832A, ICD-10-D-
	M80851A, ICD-10-D-M80851D, ICD-10-D-M80851S, ICD-10-D-

Complication	ICD-10-D Codes					
	M80852A, ICD-10-D-M80852D, ICD-10-D-M80852S, ICD-10-D- M80859A, ICD-10-D-M80861A, ICD-10-D-M80862D, ICD-10-D- M80872A, ICD-10-D-M8088XA, ICD-10-D-M8088XD, ICD-10-D- M8088XG, ICD-10-D-M8088XS, ICD-10-D-M810, ICD-10-D-M816, ICD-10-D-M818					
Psychiatric/neurologic						
Anxiety	ICD-10-D-F064, ICD-10-D-F411, ICD-10-D-F413, ICD-10-D-F418,					
Depression	ICD-10-D-F419, ICD-10-D-F4322, ICD-10-D-F4323 ICD-10-D-F320, ICD-10-D-F321, ICD-10-D-F322, ICD-10-D-F323, ICD-10-D-F324, ICD-10-D-F325, ICD-10-D-F328, ICD-10-D-F3289, ICD-10-D-F329, ICD-10-D-F330, ICD-10-D-F331, ICD-10-D-F332, ICD-10-D-F333, ICD-10-D-F3340, ICD-10-D-F3341, ICD-10-D-F3342,					
Insomnia	ICD-10-D-F338, ICD-10-D-F339, ICD-10-D-F4321, ICD-10-D-F4323 ICD-10-D-F5101, ICD-10-D-F5102, ICD-10-D-F5103, ICD-10-D- F5104, ICD-10-D-F5105, ICD-10-D-F5109, ICD-10-D-G4700, ICD-10- D-G4701, ICD-10-D-G4709, ICD-10-D-Z73810, ICD-10-D-Z73812, ICD-10-D-Z73819					
Seizure	ICD-10-D-G4089					
Renal						
Acute kidney failure	ICD-10-D-N170, ICD-10-D-N171, ICD-10-D-N172, ICD-10-D-N178, ICD-10-D-N179					
Chronic kidney disease	ICD-10-D-N181, ICD-10-D-N182, ICD-10-D-N183, ICD-10-D-N184, ICD-10-D-N185, ICD-10-D-N186, ICD-10-D-N189, ICD-10-D-N186, ICD-10-D-Z992					
Dialysis	ICD-10-D-Z4901, ICD-10-D-Z4902, ICD-10-D-Z4931, ICD-10-D-Z4932					
Focal segmental	ICD-10-D-N001, ICD-10-D-N011, ICD-10-D-N021, ICD-10-D-N031,					
glomerulosclerosis	ICD-10-D-N041, ICD-10-D-N051, ICD-10-D-N061, ICD-10-D-N071					
Hyperuricemia	ICD-10-D-E790					
Kidney hypertrophy	ICD-10-D-N2881					
Kidney stone	ICD-10-D-N132, ICD-10-D-N200, ICD-10-D-N201, ICD-10-D-N202, ICD-10-D-N209, ICD-10-D-N210, ICD-10-D-N211, ICD-10-D-N218, ICD-10-D-N219, ICD-10-D-N22, ICD-10-D-N23					
Proteinuria	ICD-10-D-R801, ICD-10-D-R809					
Severe chronic kidney disease	ICD-10-D-N184, ICD-10-D-N185, ICD-10-D-N186, ICD-10-D-Z992					
Neoplasm						
Benign neoplasm	ICD-10-D-D100, ICD-10-D-D101, ICD-10-D-D1030, ICD-10-D-D1039, ICD-10-D-D104, ICD-10-D-D105, ICD-10-D-D106, ICD-10-D-D107, ICD-10-D-D109, ICD-10-D-D110, ICD-10-D-D117, ICD-10-D-D119, ICD-10-D-D120, ICD-10-D-D121, ICD-10-D-D122, ICD-10-D-D123, ICD-10-D-D124, ICD-10-D-D125, ICD-10-D-D126, ICD-10-D-D127, ICD-10-D-D128, ICD-10-D-D129, ICD-10-D-D130, ICD-10-D-D131, ICD-10-D-D132, ICD-10-D-D1330, ICD-10-D-D1339, ICD-10-D-D134, ICD-10-D-D135, ICD-10-D-D136, ICD-10-D-D137, ICD-10-D-D139, ICD-10-D-D140, ICD-10-D-D141, ICD-10-D-D137, ICD-10-D-D1430, ICD-10-D-D1431, ICD-10-D-D1432, ICD-10-D-D144, ICD-10-D-D150, ICD-10-D-D151, ICD-10-D-D152, ICD-10-D-D159, ICD-10-D-D1600, ICD-10-D-D1601, ICD-10-D-D1602, ICD-10-D-D1612, ICD-10-D-					

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#### ICD-10-D Codes

D1620, ICD-10-D-D1621, ICD-10-D-D1622, ICD-10-D-D1631, ICD-10-D-D1632, ICD-10-D-D164, ICD-10-D-D165, ICD-10-D-D166, ICD-10-D-D167, ICD-10-D-D168, ICD-10-D-D169, ICD-10-D-D170, ICD-10-D-D171, ICD-10-D-D1720, ICD-10-D-D1721, ICD-10-D-D1722, ICD-10-D-D1723, ICD-10-D-D1724, ICD-10-D-D1730, ICD-10-D-D1739, ICD-10-D-D175, ICD-10-D-D176, ICD-10-D-D1771, ICD-10-D-D1772, ICD-10-D-D1779, ICD-10-D-D179, ICD-10-D-D1800, ICD-10-D-D1801, ICD-10-D-D1802, ICD-10-D-D1803, ICD-10-D-D1809, ICD-10-D-D181. ICD-10-D-D190. ICD-10-D-D191. ICD-10-D-D197. ICD-10-D-D200, ICD-10-D-D201, ICD-10-D-D210, ICD-10-D-D2110, ICD-10-D-D2111, ICD-10-D-D2112, ICD-10-D-D2120, ICD-10-D-D2121, ICD-10-D-D2122, ICD-10-D-D214, ICD-10-D-D215, ICD-10-D-D216, ICD-10-D-D219, ICD-10-D-D220, ICD-10-D-D2211, ICD-10-D-D2212, ICD-10-D-D22122, ICD-10-D-D2221, ICD-10-D-D2222, ICD-10-D-D2230, ICD-10-D-D2239, ICD-10-D-D224, ICD-10-D-D225, ICD-10-D-D2260, ICD-10-D-D2261, ICD-10-D-D2262, ICD-10-D-D2270, ICD-10-D-D2271, ICD-10-D-D2272, ICD-10-D-D229, ICD-10-D-D230, ICD-10-D-D2310, ICD-10-D-D2311, ICD-10-D-D23111, ICD-10-D-D23112, ICD-10-D-D2312, ICD-10-D-D23121, ICD-10-D-D23122, ICD-10-D-D2320, ICD-10-D-D2321, ICD-10-D-D2322, ICD-10-D-D2330, ICD-10-D-D2339, ICD-10-D-D234, ICD-10-D-D235, ICD-10-D-D2360, ICD-10-D-D2361, ICD-10-D-D2362, ICD-10-D-D2370, ICD-10-D-D2371, ICD-10-D-D2372, ICD-10-D-D239, ICD-10-D-D241, ICD-10-D-D242, ICD-10-D-D249, ICD-10-D-D250, ICD-10-D-D251, ICD-10-D-D252, ICD-10-D-D259, ICD-10-D-D260, ICD-10-D-D261, ICD-10-D-D267, ICD-10-D-D269, ICD-10-D-D270, ICD-10-D-D271, ICD-10-D-D279, ICD-10-D-D280, ICD-10-D-D281, ICD-10-D-D282, ICD-10-D-D287, ICD-10-D-D290. ICD-10-D-D291. ICD-10-D-D2921. ICD-10-D-D294. ICD-10-D-D3000, ICD-10-D-D3001, ICD-10-D-D3002, ICD-10-D-D3010, ICD-10-D-D303, ICD-10-D-D308, ICD-10-D-D309, ICD-10-D-D3100, ICD-10-D-D3101, ICD-10-D-D3102, ICD-10-D-D3112, ICD-10-D-D3121, ICD-10-D-D3122, ICD-10-D-D3130, ICD-10-D-D3131, ICD-10-D-D3132, ICD-10-D-D3140, ICD-10-D-D3141, ICD-10-D-D3142, ICD-10-D-D3160, ICD-10-D-D3161, ICD-10-D-D3190, ICD-10-D-D3191, ICD-10-D-D3192, ICD-10-D-D320, ICD-10-D-D321, ICD-10-D-D329, ICD-10-D-D330, ICD-10-D-D331, ICD-10-D-D332, ICD-10-D-D333, ICD-10-D-D334, ICD-10-D-D337, ICD-10-D-D339, ICD-10-D-D34, ICD-10-D-D3500, ICD-10-D-D3501, ICD-10-D-D3502, ICD-10-D-D351, ICD-10-D-D352, ICD-10-D-D360, ICD-10-D-D3610, ICD-10-D-D3612, ICD-10-D-D3613, ICD-10-D-D3614, ICD-10-D-D3615, ICD-10-D-D3616, ICD-10-D-D3617, ICD-10-D-D367, ICD-10-D-D369, ICD-10-D-D3701, ICD-10-D-D3702, ICD-10-D-D37030, ICD-10-D-D37032, ICD-10-D-D3705, ICD-10-D-D3709, ICD-10-D-D371, ICD-10-D-D372, ICD-10-D-D373, ICD-10-D-D374, ICD-10-D-D375, ICD-10-D-D376, ICD-10-D-D378, ICD-10-D-D379, ICD-10-D-D380, ICD-10-D-D381, ICD-10-D-D382, ICD-10-D-D383, ICD-10-D-D386, ICD-10-D-D390, ICD-10-D-D3910, ICD-10-D-D3911, ICD-10-D-D3912, ICD-10-D-D398, ICD-10-D-D399, ICD-10-D-D3A00, ICD-10-D-D3A012, ICD-10-D-D3A019, ICD-10-D-D3A020, ICD-10-D-D3A021, ICD-10-D-D3A026, ICD-10-D-D3A090, ICD-10-D-D3A092, ICD-10-D-D3A093, ICD-10-D-D3A098, ICD-10-D-D3A8, ICD-10-D-D400, ICD-10-D-D4010, ICD-10-D-D4011, ICD-10-D-D408. ICD-10-D-D409. ICD-10-D-D4100. ICD-10-D-D4101. ICD-10-D-D4102, ICD-10-D-D4110, ICD-10-D-D4111, ICD-10-D-D4112, ICD-10-

Complication	ICD-10-D Codes
	D-D4120, ICD-10-D-D4122, ICD-10-D-D413, ICD-10-D-D414, ICD-10-
	D-D419, ICD-10-D-D420, ICD-10-D-D429, ICD-10-D-D430, ICD-10-D-
	D432, ICD-10-D-D433, ICD-10-D-D434, ICD-10-D-D439, ICD-10-D-
	D440, ICD-10-D-D4410, ICD-10-D-D4411, ICD-10-D-D4412, ICD-10-
	D-D442, ICD-10-D-D443, ICD-10-D-D444, ICD-10-D-D447, ICD-10-D-
	D449
Hepatocellular adenoma	ICD-10-D134
Malignant neoplasm	ICD-10-D-C001, ICD-10-D-C004, ICD-10-D-C01, ICD-10-D-C020,
	ICD-10-D-C021, ICD-10-D-C022, ICD-10-D-C023, ICD-10-D-C024,
	ICD-10-D-C028, ICD-10-D-C029, ICD-10-D-C031, ICD-10-D-C039,
	ICD-10-D-C040, ICD-10-D-C048, ICD-10-D-C049, ICD-10-D-C050,
	ICD-10-D-C051, ICD-10-D-C052, ICD-10-D-C059, ICD-10-D-C060,
	ICD-10-D-C069, ICD-10-D-C07, ICD-10-D-C080, ICD-10-D-C089, ICD-10-D-C090, ICD-10-D-C091, ICD-10-D-C098, ICD-10-D-C099,
	ICD-10-D-C109, ICD-10-D-C101, ICD-10-D-C102, ICD-10-D-C108,
	ICD-10-D-C109, ICD-10-D-C101, ICD-10-D-C102, ICD-10-D-C108, ICD-10-D-C109, ICD-10-D-C111, ICD-10-D-C112, ICD-10-D-C113,
	ICD-10-D-C109, ICD-10-D-C119, ICD-10-D-C12, ICD-10-D-C130,
	ICD-10-D-C131, ICD-10-D-C132, ICD-10-D-C138, ICD-10-D-C139,
	ICD-10-D-C140, ICD-10-D-C148, ICD-10-D-C153, ICD-10-D-C154,
	ICD-10-D-C155, ICD-10-D-C158, ICD-10-D-C159, ICD-10-D-C160,
	ICD-10-D-C161, ICD-10-D-C162, ICD-10-D-C163, ICD-10-D-C165,
	ICD-10-D-C166, ICD-10-D-C168, ICD-10-D-C169, ICD-10-D-C170,
	ICD-10-D-C171, ICD-10-D-C172, ICD-10-D-C178, ICD-10-D-C179,
	ICD-10-D-C180, ICD-10-D-C181, ICD-10-D-C182, ICD-10-D-C183,
	ICD-10-D-C184, ICD-10-D-C185, ICD-10-D-C186, ICD-10-D-C187,
	ICD-10-D-C188, ICD-10-D-C189, ICD-10-D-C19, ICD-10-D-C20, ICD-
	10-D-C210, ICD-10-D-C211, ICD-10-D-C218, ICD-10-D-C220, ICD-
	10-D-C221, ICD-10-D-C222, ICD-10-D-C223, ICD-10-D-C227, ICD-
	10-D-C228, ICD-10-D-C229, ICD-10-D-C23, ICD-10-D-C240, ICD-10-
	D-C241, ICD-10-D-C248, ICD-10-D-C249, ICD-10-D-C250, ICD-10-D-
	C251, ICD-10-D-C252, ICD-10-D-C253, ICD-10-D-C254, ICD-10-D-
	C257, ICD-10-D-C258, ICD-10-D-C259, ICD-10-D-C260, ICD-10-D-
	C269, ICD-10-D-C300, ICD-10-D-C310, ICD-10-D-C311, ICD-10-D-
	C313, ICD-10-D-C318, ICD-10-D-C319, ICD-10-D-C320, ICD-10-D-
	C321, ICD-10-D-C322, ICD-10-D-C323, ICD-10-D-C328, ICD-10-D-
	C329, ICD-10-D-C33, ICD-10-D-C3400, ICD-10-D-C3401, ICD-10-D-
	C3402, ICD-10-D-C3410, ICD-10-D-C3411, ICD-10-D-C3412, ICD-10
	D-C342, ICD-10-D-C3430, ICD-10-D-C3431, ICD-10-D-C3432, ICD-
	10-D-C3480, ICD-10-D-C3481, ICD-10-D-C3482, ICD-10-D-C3490,
	ICD-10-D-C3491, ICD-10-D-C3492, ICD-10-D-C37, ICD-10-D-C381,
	ICD-10-D-C382, ICD-10-D-C383, ICD-10-D-C384, ICD-10-D-C388,
	ICD-10-D-C390, ICD-10-D-C399, ICD-10-D-C4000, ICD-10-D-C4001, ICD-10-D-C4002, ICD-10-D-C4020, ICD-10-D-C4021, ICD-10-D-
	C4022, ICD-10-D-C4032, ICD-10-D-C4020, ICD-10-D-C4021, ICD-10-D- C4022, ICD-10-D-C4032, ICD-10-D-C4080, ICD-10-D-C4090, ICD-10
	D-C410, ICD-10-D-C4032, ICD-10-D-C4080, ICD-10-D-C4090, ICD-10 D-C410, ICD-10-D-C411, ICD-10-D-C412, ICD-10-D-C413, ICD-10-D
	C414, ICD-10-D-C419, ICD-10-D-C4311, ICD-10-D-C4312, ICD-10-D
	C4330, ICD-10-D-C4339, ICD-10-D-C434, ICD-10-D-C4351, ICD-10-
	D-C4359, ICD-10-D-C4360, ICD-10-D-C4361, ICD-10-D-C4362, ICD-
	10-D-C4370, ICD-10-D-C4371, ICD-10-D-C4372, ICD-10-D-C438,
	ICD-10-D-C439, ICD-10-D-C4400, ICD-10-D-C4401, ICD-10-D-
	C4402, ICD-10-D-C44101, ICD-10-D-C441092, ICD-10-D-C44111,
	ICD-10-D-C44112, ICD-10-D-C44119, ICD-10-D-C441191, ICD-10-D-
	C441192, ICD-10-D-C44122, ICD-10-D-C44129, ICD-10-D-C44209,

Complication	ICD-10-D Codes
•	ICD-10-D-C44211, ICD-10-D-C44212, ICD-10-D-C44219, ICD-10-D-
	C44222, ICD-10-D-C44229, ICD-10-D-C44299, ICD-10-D-C44300,
	ICD-10-D-C44301, ICD-10-D-C44309, ICD-10-D-C44310, ICD-10-D-
	C44311, ICD-10-D-C44319, ICD-10-D-C44320, ICD-10-D-C44321,
	ICD-10-D-C44329, ICD-10-D-C4440, ICD-10-D-C4441, ICD-10-D-
	C4442, ICD-10-D-C44500, ICD-10-D-C44501, ICD-10-D-C44509,
	ICD-10-D-C44511, ICD-10-D-C44519, ICD-10-D-C44520, ICD-10-D-
	C44521, ICD-10-D-C44529, ICD-10-D-C44599, ICD-10-D-C44609,
	ICD-10-D-C44611, ICD-10-D-C44612, ICD-10-D-C44619, ICD-10-D-
	C44621, ICD-10-D-C44622, ICD-10-D-C44629, ICD-10-D-C44702,
	ICD-10-D-C44709, ICD-10-D-C44711, ICD-10-D-C44712, ICD-10-D-
	C44719, ICD-10-D-C44721, ICD-10-D-C44722, ICD-10-D-C44729,
	ICD-10-D-C44792, ICD-10-D-C44799, ICD-10-D-C4481, ICD-10-D-
	C4489, ICD-10-D-C4490, ICD-10-D-C4491, ICD-10-D-C4492, ICD-10-
	D-C4499, ICD-10-D-C450, ICD-10-D-C451, ICD-10-D-C452, ICD-10-
	D-C457, ICD-10-D-C459, ICD-10-D-C460, ICD-10-D-C461, ICD-10-D-
	C462, ICD-10-D-C463, ICD-10-D-C464, ICD-10-D-C4650, ICD-10-D-
	C4651, ICD-10-D-C4652, ICD-10-D-C467, ICD-10-D-C469, ICD-10-D-
	C470, ICD-10-D-C480, ICD-10-D-C481, ICD-10-D-C482, ICD-10-D-
	C488, ICD-10-D-C490, ICD-10-D-C4911, ICD-10-D-C4912, ICD-10-D-
	C4920, ICD-10-D-C4921, ICD-10-D-C4922, ICD-10-D-C493, ICD-10-
	D-C494, ICD-10-D-C495, ICD-10-D-C496, ICD-10-D-C498, ICD-10-D-C499, ICD-10-D-C49A0, ICD-10-D-C49A2, ICD-10-D-C49A3, ICD-10-
	D-C49A4, ICD-10-D-C49A9, ICD-10-D-C49A2, ICD-10-D-C49A3, ICD-10- D-C49A4, ICD-10-D-C49A9, ICD-10-D-C50011, ICD-10-D-C50012,
	ICD-10-D-C50019, ICD-10-D-C50021, ICD-10-D-C50029, ICD-10-D-
	C50111, ICD-10-D-C50112, ICD-10-D-C50119, ICD-10-D-C50121,
	ICD-10-D-C50211, ICD-10-D-C50212, ICD-10-D-C50219, ICD-10-D-
	C50222, ICD-10-D-C50311, ICD-10-D-C50312, ICD-10-D-C50319,
	ICD-10-D-C50411, ICD-10-D-C50412, ICD-10-D-C50419, ICD-10-D-
	C50511, ICD-10-D-C50512, ICD-10-D-C50519, ICD-10-D-C50611,
	ICD-10-D-C50612, ICD-10-D-C50619, ICD-10-D-C50811, ICD-10-D-
	C50812, ICD-10-D-C50819, ICD-10-D-C50829, ICD-10-D-C50911,
	ICD-10-D-C50912, ICD-10-D-C50919, ICD-10-D-C50921, ICD-10-D-
	C50922, ICD-10-D-C50929, ICD-10-D-C510, ICD-10-D-C511, ICD-10-
	D-C518, ICD-10-D-C519, ICD-10-D-C52, ICD-10-D-C530, ICD-10-D-
	C531, ICD-10-D-C538, ICD-10-D-C539, ICD-10-D-C541, ICD-10-D-
	C542, ICD-10-D-C543, ICD-10-D-C548, ICD-10-D-C549, ICD-10-D-
	C55, ICD-10-D-C561, ICD-10-D-C562, ICD-10-D-C569, ICD-10-D-
	C5700, ICD-10-D-C5701, ICD-10-D-C5702, ICD-10-D-C5710, ICD-10- D-C574, ICD-10-D-C577, ICD-10-D-C579, ICD-10-D-C600, ICD-10-D-
	C601, ICD-10-D-C602, ICD-10-D-C608, ICD-10-D-C609, ICD-10-D-
	C61, ICD-10-D-C6202, ICD-10-D-C6210, ICD-10-D-C6211, ICD-10-D-
	C6212, ICD-10-D-C6290, ICD-10-D-C6291, ICD-10-D-C6292, ICD-10-
	D-C6302, ICD-10-D-C6310, ICD-10-D-C638, ICD-10-D-C641, ICD-10-
	D-C642, ICD-10-D-C649, ICD-10-D-C651, ICD-10-D-C652, ICD-10-D-
	C659, ICD-10-D-C661, ICD-10-D-C662, ICD-10-D-C669, ICD-10-D-
	C670, ICD-10-D-C671, ICD-10-D-C672, ICD-10-D-C673, ICD-10-D-
	C674, ICD-10-D-C675, ICD-10-D-C677, ICD-10-D-C678, ICD-10-D-
	C679, ICD-10-D-C680, ICD-10-D-C688, ICD-10-D-C689, ICD-10-D-
	C6900, ICD-10-D-C6901, ICD-10-D-C6902, ICD-10-D-C6911, ICD-10-
	D-C6930, ICD-10-D-C6931, ICD-10-D-C6932, ICD-10-D-C6940, ICD-
	10-D-C6941, ICD-10-D-C6950, ICD-10-D-C6952, ICD-10-D-C6960,
	ICD-10-D-C6961, ICD-10-D-C6962, ICD-10-D-C6990, ICD-10-D-

Complication	ICD-10-D Codes
	C6991, ICD-10-D-C6992, ICD-10-D-C700, ICD-10-D-C701, ICD-10-D-
	C709, ICD-10-D-C710, ICD-10-D-C711, ICD-10-D-C712, ICD-10-D-
	C713, ICD-10-D-C714, ICD-10-D-C715, ICD-10-D-C716, ICD-10-D-
	C717, ICD-10-D-C718, ICD-10-D-C719, ICD-10-D-C720, ICD-10-D-
	C7230, ICD-10-D-C7231, ICD-10-D-C7232, ICD-10-D-C7259, ICD-10-
	D-C729, ICD-10-D-C73, ICD-10-D-C7400, ICD-10-D-C7401, ICD-10-
	D-C7402, ICD-10-D-C7412, ICD-10-D-C7490, ICD-10-D-C7491, ICD-
	10-D-C7492, ICD-10-D-C750, ICD-10-D-C751, ICD-10-D-C752, ICD-
	10-D-C753, ICD-10-D-C755, ICD-10-D-C758, ICD-10-D-C759, ICD-
	10-D-C760, ICD-10-D-C761, ICD-10-D-C762, ICD-10-D-C763, ICD-
	10-D-C7642, ICD-10-D-C7650, ICD-10-D-C7651, ICD-10-D-C7652,
	ICD-10-D-C768, ICD-10-D-C770, ICD-10-D-C771, ICD-10-D-C772,
	ICD-10-D-C773, ICD-10-D-C774, ICD-10-D-C775, ICD-10-D-C778,
	ICD-10-D-C779, ICD-10-D-C7800, ICD-10-D-C7801, ICD-10-D-
	C7802, ICD-10-D-C781, ICD-10-D-C782, ICD-10-D-C7830, ICD-10-D-
	C7839, ICD-10-D-C784, ICD-10-D-C785, ICD-10-D-C786, ICD-10-D-
	C787, ICD-10-D-C7880, ICD-10-D-C7889, ICD-10-D-C7900, ICD-10-
	D-C7901, ICD-10-D-C7902, ICD-10-D-C7910, ICD-10-D-C7911, ICD-
	10-D-C7919, ICD-10-D-C792, ICD-10-D-C7931, ICD-10-D-C7932,
	ICD-10-D-C7940, ICD-10-D-C7949, ICD-10-D-C7951, ICD-10-D-
	C7952, ICD-10-D-C7960, ICD-10-D-C7961, ICD-10-D-C7962, ICD-10-
	D-C7970, ICD-10-D-C7971, ICD-10-D-C7972, ICD-10-D-C7981, ICD-
	10-D-C7982, ICD-10-D-C7989, ICD-10-D-C799, ICD-10-D-C7A00,
	ICD-10-D-C7A010, ICD-10-D-C7A012, ICD-10-D-C7A019, ICD-10-D-
	C7A020, ICD-10-D-C7A021, ICD-10-D-C7A022, ICD-10-D-C7A025,
	ICD-10-D-C7A026, ICD-10-D-C7A029, ICD-10-D-C7A090, ICD-10-D-
	C7A091, ICD-10-D-C7A092, ICD-10-D-C7A093, ICD-10-D-C7A094,
	ICD-10-D-C7A095, ICD-10-D-C7A098, ICD-10-D-C7A1, ICD-10-D-
	C7A8, ICD-10-D-C7B00, ICD-10-D-C7B01, ICD-10-D-C7B02, ICD-10-
	D-C7B03, ICD-10-D-C7B04, ICD-10-D-C7B09, ICD-10-D-C7B8, ICD-
	10-D-C800, ICD-10-D-C801, ICD-10-D-C802, ICD-10-D-D0000, ICD-
	10-D-D0003, ICD-10-D-D0004, ICD-10-D-D0005, ICD-10-D-D0006,
	ICD-10-D-D0007, ICD-10-D-D0008, ICD-10-D-D002, ICD-10-D-D010,
	ICD-10-D-D011, ICD-10-D-D012, ICD-10-D-D013, ICD-10-D-D015,
	ICD-10-D-D017, ICD-10-D-D020, ICD-10-D-D0220, ICD-10-D-D0221,
	ICD-10-D0222, ICD-10-D-00339, ICD-10-D-0034, ICD-10-D-
	D0352, ICD-10-D-D0359, ICD-10-D-D0361, ICD-10-D-D0362, ICD-10-
	D-D0371, ICD-10-D-D0372, ICD-10-D-D038, ICD-10-D-D039, ICD-10-
	D-D040, ICD-10-D-D0411, ICD-10-D-D0412, ICD-10-D-D0421, ICD-
	10-D-D0422, ICD-10-D-D0430, ICD-10-D-D0439, ICD-10-D-D044,
	ICD-10-D-D045, ICD-10-D-D0460, ICD-10-D-D0461, ICD-10-D-
	D0462, ICD-10-D-D0470, ICD-10-D-D0471, ICD-10-D-D0472, ICD-10-
	D-D048, ICD-10-D-D049, ICD-10-D-D0501, ICD-10-D-D0502, ICD-10-
	D-D0510, ICD-10-D-D0511, ICD-10-D-D0512, ICD-10-D-D0580, ICD-
	10-D-D0581, ICD-10-D-D0582, ICD-10-D-D0590, ICD-10-D-D0591,
	ICD-10-D-D0592, ICD-10-D-D060, ICD-10-D-D067, ICD-10-D-D069,
	ICD-10-D-D070, ICD-10-D-D071, ICD-10-D-D0730, ICD-10-D-D0739,
	ICD-10-D-D075, ICD-10-D-D090, ICD-10-D-D0910, ICD-10-D-D0919,
	ICD-10-D-D0922, ICD-10-D-D098, ICD-10-D-D099
Primary liver cancer	ICD-10-D-C220, ICD-10-D-C221, ICD-10-D-C222, ICD-10-D-C223,
	ICD-10-D-C224, ICD-10-D-C227, ICD-10-D-C228, ICD-10-D-C229
Liver and/or kidney transplant	

Complication	ICD-10-D Codes
Liver transplant	ICD-10-D-T8640, ICD-10-D-T8641, ICD-10-D-T8642, ICD-10-D-
	T8643, ICD-10-D-T8649, ICD-10-D-Z4823, ICD-10-D-Z944
Kidney transplant	ICD-10-D-T8610, ICD-10-D-T8611, ICD-10-D-T8612, ICD-10-D-
	T8613, ICD-10-D-T8619, ICD-10-D-Z4822, ICD-10-D-Z940

Abbreviations: GSDIa, glycogen storage disease type Ia; ICD-10-CM, International Classification of Diseases, Tenth Revision–Clinical Modification.

# Table S2. Prevalence of Complications

	Pediatric		Adult		Total	
	GSDIa (n=185)	Comparator (n=1850)	GSDIa (n=372)	Comparator (n=3720)	GSDIa (n=557)	Comparator (n=5570)
Any	167 (90.3)	815 (44.1)	356 (95.7)	2737 (73.6)	523 (93.9)	3552 (63.8)
Kidney and/or liver transplant	NR <sup>a</sup>	0	12 (3.2)	NR <sup>a</sup>	17 (3.1)	NR <sup>a</sup>
Liver transplant	NRª	0	11 (3.0)	$NR^{a}$	16 (2.9)	NR <sup>a</sup>
Kidney transplant	NRª	0	NRª	NR	NR <sup>a</sup>	$NR^{a}$
Hematologic	117 (63.2)	49 (2.7)	230 (61.8)	329 (8.8)	347 (62.3)	378 (6.8)
Acidosis	37 (20.0)	NRª	64 (17.2)	24 (0.7)	101 (18.1)	27 (0.5)
Anemia due to enzyme disorders	81 (43.8)	0	151 (40.6)	NRª	232 (41.7)	$NR^{a}$
Anemia	42 (22.7)	47 (2.5)	138 (37.1)	321 (8.6)	180 (32.3)	368 (6.6)
Iron deficiency anemia	22 (11.9)	16 (0.9)	62 (16.7)	127 (3.4)	84 (15.1)	143 (2.6)
Hepatic	71 (38.4)	54 (2.9)	204 (54.8)	1074 (28.9)	275 (49.4)	1128 (20.3)
Hepatomegaly	53 (28.7)	NRª	59 (15.9)	28 (0.8)	112 (20.1)	32 (0.6)
Hypoglycemia	50 (27.0)	22 (1.2)	42 (11.3)	32 (0.9)	92 (16.5)	54 (1.0)
Hyperlipidemia	20 (10.8)	28 (1.5)	168 (45.2)	1047 (28.2)	188 (33.8)	1075 (19.3)
Renal	33 (17.8)	12 (0.7)	120 (32.3)	248 (6.7)	153 (27.5)	260 (4.7)
Hyperuricemia	NRª	0	19 (5.1)	12 (0.3)	27 (4.9)	12 (0.2)
Dialysis	0	0	NRª	$NR^{a}$	NRª	NRª
Kidney hypertrophy	NRª	0	$NR^{a}$	0	$NR^{a}$	0
Acute kidney failure	NR <sup>a</sup>	NR <sup>a</sup>	57 (15.3)	54 (1.5)	62 (11.1)	55 (1.0)

	Pediatric		Adult		Total	
	GSDIa (n=185)	Comparator (n=1850)	GSDla (n=372)	Comparator (n=3720)	GSDIa (n=557)	Comparator (n=5570)
Proteinuria	NR <sup>a</sup>	NR <sup>a</sup>	21 (5.7)	31 (0.8)	28 (5.0)	38 (0.7)
Chronic kidney disease	$NR^{a}$	NR <sup>a</sup>	48 (12.9)	88 (2.4)	55 (9.9)	90 (1.6)
Severe chronic kidney disease	NR <sup>a</sup>	0	16 (4.3)	19 (0.5)	17 (3.1)	19 (0.3)
Kidney stone	14 (7.6)	NRª	42 (11.3)	121 (3.3)	56 (10.1)	123 (2.2)
Focal segmental glomerulosclerosis	0	0	NR <sup>a</sup>	$NR^{a}$	NR <sup>a</sup>	NR
Nutritional	114 (61.6)	222 (12.0)	221 (59.4)	1152 (31.0)	335 (60.1)	1374 (24.7)
Gastrostomy	55 (29.7)	NRª	12 (3.2)	$NR^{a}$	67 (12.0)	$NR^{a}$
Poor growth	41 (22.2)	70 (3.8)	NR <sup>a</sup>	$NR^{a}$	48 (8.6)	75 (1.4)
Anorexia	NRª	NRª	18 (4.8)	15 (0.4)	26 (4.7)	25 (0.5)
Nutritional deficiency	59 (31.9)	44 (2.4)	150 (40.3)	594 (16.0)	209 (37.5)	638 (11.5)
Protein-calorie malnutrition	$NR^{a}$	NR <sup>a</sup>	18 (4.8)	24 (0.7)	26 (4.7)	26 (0.5)
Iron deficiency	29 (15.7)	19 (1.0)	65 (17.5)	137 (3.7)	94 (16.9)	156 (2.8)
Vitamin D deficiency	33 (17.8)	20 (1.1)	90 (24.2)	458 (12.3)	123 (22.1)	478 (8.6)
Other nutritional deficiency	$NR^{a}$	NR <sup>a</sup>	27 (7.3)	137 (3.7)	31 (5.6)	145 (2.6)
Obesity	35 (18.9)	114 (6.2)	116 (31.2)	718 (19.3)	151 (27.1)	832 (14.9)
Digestive system	104 (56.2)	461 (24.9)	241 (64.8)	1173 (31.5)	345 (61.9)	1634 (29.3)
Pancreatitis	NRª	0	11 (3.0)	11 (0.3)	12 (2.2)	11 (0.2)
Intestinal gas	NR <sup>a</sup>	20 (1.1)	38 (10.2)	95 (2.6)	48 (8.6)	115 (2.1)
Nausea and/or vomiting	81 (43.8)	235 (12.7)	133 (35.8)	340 (9.1)	214 (38.4)	575 (10.3)

	Pediatric		Adult		Total	
	GSDIa (n=185)	Comparator (n=1850)	GSDIa (n=372)	Comparator (n=3720)	GSDIa (n=557)	Comparator (n=5570)
Gallstones	NR <sup>a</sup>	0	29 (7.8)	72 (1.9)	32 (5.8)	72 (1.3)
Diarrhea	49 (26.5)	130 (7.0)	69 (18.6)	212 (5.7)	118 (21.2)	342 (6.1)
Abdominal pain	51 (27.6)	198 (10.7)	162 (43.6)	718 (19.3)	213 (38.2)	916 (16.5)
Gastroesophageal reflux disease	26 (14.1)	97 (5.2)	117 (31.5)	487 (13.1)	143 (25.7)	584 (10.5)
Musculoskeletal	36 (19.5)	96 (5.2)	202 (54.3)	1036 (27.9)	238 (42.7)	1132 (20.3)
Gout	NR <sup>a</sup>	0	44 (11.8)	81 (2.2)	45 (8.1)	81 (1.5)
Osteoporosis	NR <sup>a</sup>	NR <sup>a</sup>	27 (7.3)	76 (2.0)	32 (5.8)	77 (1.4)
Fatigue	30 (16.2)	92 (5.0)	130 (35.0)	726 (19.5)	160 (28.7)	818 (14.7)
Osteoarthritis	NR <sup>a</sup>	NRª	82 (22.0)	379 (10.2)	87 (15.6)	383 (6.9)
Cardiovascular/cardiopulmonary	29 (15.7)	16 (0.9)	207 (55.7)	1045 (28.1)	236 (42.4)	1061 (19.1)
Pulmonary hypertension	0	NRª	11 (3.0)	16 (0.4)	11 (2.0)	18 (0.3)
Hypertension	16 (8.7)	NRª	185 (49.7)	1009 (27.1)	201 (36.1)	1015 (18.2)
Atherosclerotic heart disease	0	NRª	32 (8.6)	157 (4.2)	32 (5.8)	158 (2.8)
Neoplasm	18 (9.7)	104 (5.6)	153 (41.1)	842 (22.6)	171 (30.7)	946 (17.0)
Malignant neoplasm	NR <sup>a</sup>	NRª	40 (10.8)	197 (5.3)	43 (7.7)	199 (3.6)
Primary liver cancer	0	0	NRª	0	NRª	0
Benign neoplasm	15 (8.1)	102 (5.5)	139 (37.4)	772 (20.8)	154 (27.7)	874 (15.7)
Hepatocellular adenoma	NR <sup>a</sup>	0	26 (7.0)	$NR^{a}$	29 (5.2)	$NR^{a}$
Psychiatric/neurologic	24 (13.0)	176 (9.5)	143 (38.4)	1004 (27.0)	167 (30.0)	1180 (21.2)

	Pediatric		Adult		Total	
	GSDla (n=185)	Comparator (n=1850)	GSDIa (n=372)	Comparator (n=3720)	GSDla (n=557)	Comparator (n=5570)
Seizure	NR <sup>a</sup>	NR <sup>a</sup>	NR <sup>a</sup>	15 (0.4)	NR <sup>a</sup>	18 (0.3)
Depression	12 (6.5)	74 (4.0)	89 (23.9)	540 (14.5)	101 (18.1)	614 (11.0)
Insomnia	$NR^{a}$	22 (1.2)	37 (10.0)	223 (6.0)	39 (7.0)	245 (4.4)
Anxiety	18 (9.7)	139 (7.5)	98 (26.3)	715 (19.2)	116 (20.8)	854 (15.3)

Abbreviations: GSDIa, glycogen storage disease type Ia; NR, not reported.

<sup>a</sup>Patient counts of 1 to 10 are not reported for privacy reasons.

 Table S3. Possible Complication Etiologies

Complication	Possible Etiology			
Digestive system				
Abdominal pain	Large amounts of raw cornstarch have been found to cause abdo pain; gallstones and pancreatitis may also contribute			
Nausea and/or vomiting	Need for emergency care if oral feeds are not tolerated			
Gastroesophageal reflux disease	Obesity and anxiety may contribute			
Diarrhea	Large amounts of raw cornstarch can modify microbiota leading to diarrhea			
Gastrostomy	Cornstarch and dietary management (continuous/nocturnal feeding)			
Intestinal gas	Large amounts of raw cornstarch have been found to cause flatulence			
Gallstones	Hyperlipidemia			
Pancreatitis	Hyperlipidemia			
Nutritional				
Nutritional deficiency	Dietary restrictions, management with cornstarch, digestive complications			
Vitamin D deficiency	Dietary restriction of lactose and galactose			
Iron deficiency	Cornstarch reduces iron bioavailability in the gastrointestinal tract			
Protein-calorie malnutrition	Anorexia, liver and/or kidney disease, and cancer can result in disturbances of protein intake/metabolism			
Other	Dietary restrictions, management with cornstarch, and digestive complications			
Poor growth	Hypoglycemia, nutritional deficiency, and chronic acidosis			
Obesity	Consumption of large amounts of raw cornstarch which is high in calories and reduced exercise capacity			
Anorexia	Need for emergency care if unable to adhere to dietary management			
Hematologic				
Anemia due to enzyme disorders	Possible miscoding or upcoding, as GSDIa is an enzyme disorder and anemia is a common complication			
Anemia	Decreased production of erythropoietin due to renal insufficiency, chronic lactic acidosis, iron deficiency, and complications of hepatic adenoma			
Iron deficiency anemia	Iron deficiency and bleeding due to altered platelet function			
Acidosis	Progressive renal disease and lactic acidosis caused by repeated hypoglycemia			
Hepatic				
Hepatomegaly	Accumulation of glucose in the liver			
Hyperlipidemia	Increased lipid synthesis and decreased lipid serum clearance			

Hypoglycemia	Deficiency of glucose-6-phosphatase results in impaired release of glucose from glycogen		
Cardiovascular/cardiopulmonary			
Hypertension	Progressive renal disease		
Atherosclerotic heart disease	Hypertension, hyperlipidemia, and obesity		
Pulmonary hypertension	Abnormal production of serotonin, a vasoconstrictive amine (hypothesis)		
Musculoskeletal			
Fatigue	Nutritional deficiency and hypoglycemia		
Osteoarthritis	Obesity, gout, and reduced exercise capacity		
Gout	Hyperuricemia stemming from altered renal tubular function		
Osteoporosis	Lactic acidosis and cortisol release secondary to hypoglycemia and vitamin D deficiency and inadequate calcium intake due to dietary restrictions		
Psychiatric/neurologic			
Anxiety	Diagnosis of rare disease and highest emotional burden in those with a rare disease where there are no available treatments		
Depression	Diagnosis of rare disease and highest emotional burden in those with a rare disease where there are no available treatments		
Insomnia	Ingestion of raw cornstarch every 4-5 hours including at night and recommended to prevent hypoglycemia in adults		
Seizure	Hypoglycemia		
Renal			
Acute kidney failure Chronic kidney disease Dialysis Focal segmental glomerulosclerosis	Activation of the renin–angiotensin system, prolonged oxidative streat and profibrotic cytokines may be implicated in glomerular injury and progressive renal disease		
Proteinuria			
Kidney hypertrophy	Accumulation of glycogen and fat in the kidneys		
Kidney stone	Chronic acidosis, hypocitraturia, hypercalciuria, and hyperuricemia are thought to be contributing factors		
Hyperuricemia	Hyperuricemia due to altered renal tubular function		
Neoplasm			
Malignant neoplasm			
Primary liver cancer	Etiology of hepatocellular adenomas and transformation to hepatocellular carcinoma is multifactorial; insufficient metabolic control may play a role		
Benign neoplasm			

Hepatocellular adenoma	Etiology of hepatocellular adenomas and transformation to hepatocellular carcinoma is multifactorial; insufficient metabolic con appears to play a role	
Liver and/or kidney transplant		
Liver transplant	Accumulation of glycogen and fat in the liver, leading to progressive organ failure and tumors (adenomas and malignancies); may be preemptive to address underlying disease	
Kidney transplant	Accumulation of glycogen and fat in the kidneys, leading to progressive organ failure	

## Figure S1. Complications in Children With GSDIa vs Comparators

	GSDIa, n (%)	Comparator, n (%)		OR (95% CI)
Hematologic	117 (63.2)	49 (2.7)	H	63.2 (41.9–95.5)
Acidosis	37 (20)	NR*	⊢-●1	153.9 (46.9–505.0)
Anemia	42 (22.7)	47 (2.5)	H	11.3 (7.2–17.7)
Iron deficiency anemia	22 (11.9)	16 (0.9)	⊢●⊣	15.5 (8.0–30.0)
Renal	33 (17.8)	12 (0.7)	<b>⊢</b> •−1	33.3 (16.8–65.7)
Kidney stone	14 (7.6)	NR*	<b>⊢</b> +	75.7 (17.1–335.6)
Cardiovascular/cardiopulmonary	29 (15.7)	16 (0.9)	<b>⊢</b> •-1	21.3 (11.3–40.1)
Hypertension	16 (8.7)	NR*	⊢●1	29.1 (11.2–75.3)
Hepatic	71 (38.3)	54 (2.9)	н	20.7 (13.9–31.0)
Hepatomegaly	53 (28.7)	NR*	<b>⊢</b>	185.3 (66.0–519.7)
Hypoglycemia	50 (27.0)	22 (1.2)	⊢●⊣	30.8 (18.1–52.3)
Hyperlipidemia	20 (10.8)	28 (1.5)	⊢●⊣	7.9 (4.4–14.3)
Nutritional	114 (61.6)	222 (12.0)	ю	11.8 (8.5–16.3)
Gastrostomy	55 (29.7)	NR*	<b>⊢</b> ●1	195.2 (69.7–546.9)
Poor growth	41 (21.2)	70 (3.8)	H	7.2 (4.8–11.0)
Nutritional deficiency	59 (31.9)	44 (2.4)	HeH	19.2 (12.5–29.6)
Iron deficiency	29 (15.7)	19 (1.0)	⊢●⊣	17.9 (9.8–32.7)
Vitamin D deficiency	33 (17.8)	20 (1.1)	⊢●┥	19.9 (11.1–35.5)
Obesity	35 (18.9)	114 (6.2)	HeH	3.6 (2.4-5.4)
Musculoskeletal	36 (19.5)	96 (5.2)	H	4.4 (2.9-6.7)
Fatigue	30 (16.2)	92 (5.0)	H	3.7 (2.4–5.8)
Digestive system	104 (56.2)	461 (24.9)	H	3.9 (2.8–5.3)
Intestinal gas	NR*	20 (1.1)	⊢●⊣	5.2 (2.4–11.4)
Nausea and/or vomiting	81 (43.8)	235 (12.7)	IOI	5.4 (3.9-7.4)
Diarrhea	49 (26.5)	130 (7.0)	H€H	4.8 (3.3-6.9)
Abdominal pain	51 (27.6)	198 (10.7)	Hel	3.2 (2.2-4.5)
GERD	26 (14.1)	97 (5.2)	HeH	3.0 (1.9–4.7)
Neoplasm	18 (9.7)	104 (5.6)	⊢●⊣	1.8 (1.1–3.1)
Benign neoplasm	15 (8.1)	102 (5.5)	H <b>●</b> -I	1.5 (0.9–2.7)
Psychiatric/neurologic	24 (13.0)	176 (9.5)	HOH	1.4 (0.9–2.2)
Depression	12 (6.5)	74 (4.0)	⊢●-1	1.7 (0.9–3.1)
Insomnia	NR*	22 (1.2) ⊢	<b></b> ●I	0.9 (0.2–3.9)
Anxiety	18 (9.7)	139 (7.5)		1.3 (0.8–2.2)
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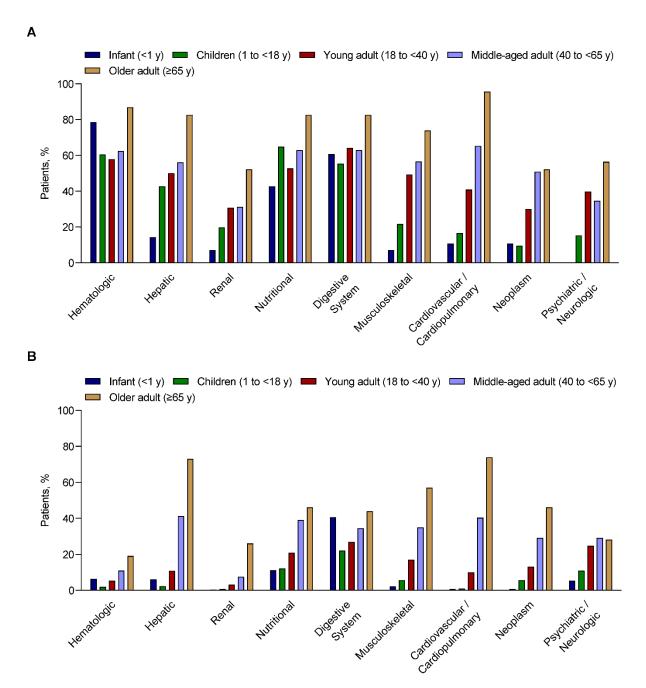
Abbreviations: GSDIa, glycogen storage disease type Ia; CI, confidence interval; NR, not reported; OR, odds ratio. \*Patient counts of 1 to 10 were not reported for privacy reasons.

#### Figure S2. Complications in Adults With GSDIa vs Comparators

	GSDIa, n (%)	Comparator, n (%)		OR (95% CI)
Liver and/or kidney transplant	12 (3.2)	NR*	<b>⊢</b> ∎–1	41.3 (11.6–147.0)
Liver transplant	11 (3.0)	NR*	<b>⊢_</b> ●	113.3 (14.6–880.1)
Hematologic	230 (61.8)	329 (8.8)	M	16.7 (13.2-21.2)
Acidosis	64 (17.2)	24 (0.7)	HEH	32.0 (19.7–51.9)
Anemia	138 (37.1)	321 (8.6)		6.3 (4.9–7.9)
Iron deficiency anemia	62 (16.7)	127 (3.4)	lei	5.7 (4.1-7.8)
Renal	120 (32.3)	248 (6.7)	101	6.7 (5.2-8.6)
Hyperuricemia	19 (5.1)	12 (0.3)	H	16.6 (8.0–34.5)
Acute kidney failure	57 (15.3)	54 (1.5)	Hei	12.3 (8.3–18.1)
Proteinuria	21 (5.7)	31 (0.8)	H€H	7.1 (4.1–12.5)
Chronic kidney disease	48 (12.9)	88 (2.4)	Hei	6.1 (4.2-8.9)
Severe chronic kidney disease	16 (4.3)	19 (0.5)	⊢●⊣	8.8 (4.5–17.2)
Kidney stone	42 (11.3)	121 (3.3)	Hei	3.8 (2.6–5.5)
Digestive system	241 (64.8)	1173 (31.5)		4.0 (3.2-5.0)
Pancreatitis	11 (3.0)	11 (0.3)	⊢●┥	10.3 (4.4–23.9)
Intestinal gas	38 (10.2)	95 (2.6)	Hei	4.3 (2.9–6.4)
Nausea and/or vomiting	133 (35.8)	340 (9.1)		5.5 (4.4-7.0)
Gallstones	29 (7.8)	72 (1.9)	IOI	4.3 (2.8–6.7)
Diarrhea	69 (18.6)	212 (5.7)		3.8 (2.8–5.1)
Abdominal pain	162 (43.6)	718 (19.3)	M	3.2 (2.6–4.0)
GERD	117 (31.5)	487 (13.1)		3.1 (2.4–3.9)
Nutritional	221 (59.4)	1152 (31.0)		3.3 (2.6–4.1)
Gastrostomy	12 (3.2)	NR*	<b>⊢</b>	124.0 (16.1–955.9)
Anorexia	18 (4.8)	15 (0.4)	⊢●⊣	12.6 (6.3–25.1)
Nutritional deficiency	150 (40.3)	594 (16.0)		3.6 (2.8–4.5)
Protein-calorie malnutrition	18 (4.8)	24 (0.7)	HeH	7.8 (4.2–14.6)
Iron deficiency	65 (17.5)	137 (3.7)	H	5.5 (4.0–7.6)
Vitamin D deficiency	90 (24.2)	458 (12.3)		2.3 (1.8–2.9)
Other nutritional deficiency	27 (7.3)	137 (3.7)	Hel	2.1 (1.3–3.1)
Obesity	116 (31.2)	718 (19.3)		1.9 (1.5–2.4)
Cardiovascular/cardiopulmonary	207 (55.7)	1045 (28.1)		3.2 (2.6–4.0)
Pulmonary hypertension	11 (3.0)	16 (0.4)	⊢●⊣	7.1 (3.3–15.3)
Hypertension	185 (49.7)	1009 (27.1)		2.7 (2.1–3.3)
Atherosclerotic heart disease	32 (8.6)	157 (4.2)	H	2.1 (1.4–3.2)
Musculoskeletal	202 (54.3)	1036 (27.9)		3.1 (2.5–3.8)
Gout	44 (11.8)	81 (2.2)	H	6.0 (4.1-8.9)
Osteoporosis	27 (7.3)	76 (2.0)	Hei	3.8 (2.4–5.9)
Fatigue	130 (35.0)	726 (19.5)	M	2.2 (1.8–2.8)
Osteoarthritis	82 (22.0)	379 (10.2)		2.5 (1.9–3.3)
Hepatic	204 (54.8)	1074 (28.9)		3.0 (2.4-3.7)
Hepatomegaly	59 (15.9)	28 (0.8)	Heri	24.9 (15.6–39.5)
Hypoglycemia	42 (11.3)	32 (0.9)	н <del>ө</del> н	14.7 (9.1–23.6)
Hyperlipidemia	168 (45.2)	1047 (28.2)		2.1 (1.7–2.6)
Neoplasm	153 (41.1)	842 (22.6)		2.4 (1.9–3.0)
Malignant neoplasm	40 (10.8)	197 (5.3)	l <del>o</del> l	2.2 (1.5–3.1)
Benign neoplasm	139 (37.3)	772 (20.8)		2.3 (1.8–2.9)
Hepatocellular adenoma	26 (7.0)	NR*	<b>⊢</b> •i	279.5 (37.8–2065.6)
Psychiatric/neurologic	143 (38.4)	1004 (27.0)		1.7 (1.4–2.1)
Seizure	NR*	15 (0.4)	<b>⊢</b> ●1	1.3 (0.3–5.9)
Depression	89 (23.9)	540 (14.5)		1.9 (1.4–2.4)
Insomnia	37 (10.0)	223 (6.0)	lei	1.7 (1.2–2.5)
Anxiety	98 (26.3)	715 (19.2)		1.5 (1.2–1.9)
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Abbreviations: CI, confidence interval; GSDIa, glycogen storage disease type Ia; NR, not reported; OR, odds ratio.

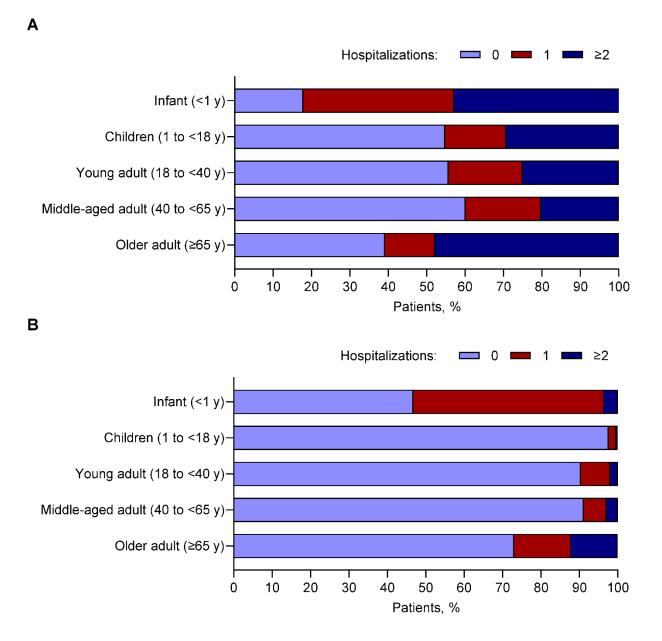
\*Patient counts of 1-10 were not reported for privacy reasons.



**Figure S3.** Frequency of Complication Types by Patient Age in Patients With GSDIa (**A**) and in Comparators (**B**)

Abbreviation: GSDIa, glycogen storage disease type Ia.

**Figure S4.** Frequency of Hospitalizations by Patient Age in Patients With GSDIa (**A**) and in Comparators (**B**)



Abbreviation: GSDIa, glycogen storage disease type Ia.