

n°	Orpha ID	Rare diseases	ICD-10	Titles of ICD blocks
1	855	Hashimoto struma	E06.3	Endocrine, nutritional and metabolic diseases
2	178029	Central diabetes insipidus	E23.2	
3	99889	Ectopic Cushing syndrome	E24.3	
4	3453	Autoimmune polyendocrinopathy type 1	E31.0	
5	355	Gaucher disease	E75.2	
6	77260	Gaucher disease, type 2	E75.2	
7	77261	Gaucher disease, type 3	E75.2	
8	85212	Perinatal-lethal Gaucher disease	E75.2	
9	530	Lipoid proteinosis	E75.5	
10	565	Menkes disease	E83.0	
11	905	Wilson disease	E83.0	
12	183	Churg-Strauss syndrome	M30.1	
13	2331	Kawasaki disease	M30.3	
14	93585	Acquired thrombotic thrombocytopenic purpura due to anti-ADAMTS 13	M31.3	
15	536	Systemic lupus erythematosus	M32	
16	801	Scleroderma	M34	
17	117	Behcet disease	M35.2	
18	49041	Retroperitoneal fibrosis	M35.5	
19	337	Fibrodysplasia ossificans progressiva	M61.1	
20	2778	Juvenile chronic recurrent multifocal osteomyelitis	M86.3	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism
21	2134	Atypical hemolytic uremic syndrome	D58.8	
22	90038	Typical hemolytic uremic syndrome	D59.3	
23	447	Paroxysmal nocturnal hemoglobinuria	D59.5	
24	903	Von Willebrand disease	D68.0	
25	54057	Thrombotic thrombocytopenic purpura	D69.4	
26	51636	WHIM syndrome	D81.0	
27	797	Sarcoidosis	D86	Diseases of the circulatory system
28	422	Idiopathic and/or familial pulmonary arterial hypertension	I27.0	
29	136	CADASIL syndrome*	I67.3	
30	2573	Moyamoya disease	I67.5	
31	36258	Buerger's disease	I73.1	
32	131	Budd-Chiari syndrome	I82.0	
33	538	Lymphangioliomyomatosis	I89.8	Diseases of the eye and adnexa
34	179	Chorioretinopathy, Birdshot type	H30.9	
35	1571	Knobloch syndrome	H33.5	
36	190	Coats disease	H35.0	
37	40923	Eales disease	H35.0	
38	90050	Retinopathy of prematurity	H35.1	Congenital malformations, deformations and chromosomal abnormalities
39	649	Norrie disease	Q15.8	
41	2092	Focal dermal hypoplasia	Q82.8	
42	758	Pseudoxanthoma elasticum	Q82.8	
43	206	Crohn disease	K50	Diseases of the digestive system
44	2137	Chronic autoimmune hepatitis	K75.4	
45	803	Amyotrophic lateral sclerosis	G12.2	Diseases of the nervous system
46	563	Peripartum cardiomyopathy	O90.3	Pregnancy, childbirth and the puerperium
47	2346	Angio-osteohypertrophic syndrome*	D18.0	Neoplasms
48	79126	Acute interstitial pneumonia	NONE	No ICD classification
49	279	Age-related macular degeneration	NONE	
50	160	Castleman disease	NONE	
51	98044	Central nervous system malformation	NONE	
52	2041	Coronary arterial fistulas	NONE	
53	137698	Cytomegalovirus disease in patients with impaired cell mediated immunity deemed at risk	NONE	
54	97339	Dural sinus malformation	NONE	
55	199323	Endophthalmitis	NONE	
56	221126	Fowler syndrome	NONE	
57	95509	Granulomatous hypophysitis	NONE	
58	158032	Hemophagocytic syndrome	NONE	
59	85458	Hereditary cerebral hemorrhage with amyloidosis	NONE	
60	100006	Hereditary cerebral hemorrhage with amyloidosis, Dutch type	NONE	
61	69665	Intrahepatic cholestasis of pregnancy	NONE	
62	93448	Lysosomal storage disease with skeletal involvement	NONE	
63	101338	Mediterranean spotted fever	NONE	
64	54370	Membranoproliferative glomerulonephritis	NONE	
65	93686	Multicentric Castleman disease	NONE	
66	94058	Neovascular glaucoma	NONE	
67	182090	Pulmonary arterial hypertension	NONE	
68	199241	Pulmonary capillary hemangiomatosis	NONE	
69	71198	Rare pulmonary hypertension	NONE	
70	60032	Recurrent respiratory papillomatosis	NONE	
71	102021	Rickettsiae disease	NONE	
72	36426	Stevens-Johnson syndrome	NONE	
73	3243	Sweet syndrome	NONE	
74	90291	Systemic sclerosis	NONE	
75	93573	Thrombotic microangiopathy	NONE	
76	52759	Vasculitis	NONE	
77	98668	Vitreoretinopathy	NONE	

\*CADASIL syndrome [ORPHA136] and Angio-osteohypertrophic syndrome [ORPHA2346] have also F01.1 (Mental and behavioural disorders) and M85.8 (Diseases of the musculoskeletal system and connective tissue) ICD codes respectively.