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REVIEW ARTICLE

Classification of Rare Cardiovascular Diseases (RCD Classification), Krakow 2013

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Almost every day brings new reports of a "rare" ("orphan") disease – a disease that requires multidisciplinary knowledge and particular caution in making diagnostic and therapeutic decisions.

The Classification of Rare Cardiovascular Diseases (RCD) is aimed at (1) facilitating recognition of RCDs, and (2) grouping the expertise in the main fields of RCDs.

The classification provides a systemic framework for clinical examples selected from a broad group of patients who have been consulted in the CRCD on a regular basis by national and international experts during live videoconferences.

RCD Classification presented in Table 1 and published simultaneously in the *Journal of Rare Cardiovascular Diseases* is based on the CRCD experience of over 300 patients consulted in the years 2006–2013 (many of whom were diagnosed and treated in the CRCD) and takes into consideration the majority of publications available through PubMed. RCD Classification encompasses the diseases whose major pathological mechanism affects the cardiovascular system.

RCD Classification accommodates the intensity of clinical symptoms and pathology concerning the systemic and pulmonary circulation (Class I and Class II), the heart and myocardium (Class III), congenital heart diseases (class IV), and rhythm and conduction disorders (Class V). Cardiovascular diseases in oncological patients (Class VI) and those in pregnant patients are classified separately (Class VI and Class VII, respectively). There are also overlapping syndromes and diseases that cannot be unequivocally classified into any of the Classes I to VII, that is represented by Class VIII.

The main classes of the RCD classification include:

- Class I – rare diseases of systemic circulation
- Class II – rare diseases of pulmonary circulation
- Class III – rare diseases of the heart (cardiomyopathies)
- Class IV – rare congenital cardiovascular diseases
- Class V – rare arrhythmias
- Class VI – cardiac tumors and cardiovascular diseases in malignancy
- Class VII – cardiovascular diseases in pregnancy
- Class VIII – unclassified rare cardiovascular diseases

RCD Classification is presented in Table 1.

It is listed by groups and subgroups as appropriate. It contains RCD Classification code and the code of International Classification of Diseases (ICD-10). The main classes are arranged in order from class I to class VIII. Each entity or group of entities is assigned a unique RCD Classification code. Coding of Class VII is described in a relevant commentary in the Table 1. Consecutive unclassified rare cardiovascular cases included in Class VIII are assigned subsequent code according to the order of publication on the CRCD webpage – www.crcd.eu or in the *Journal of Rare Cardiovascular Diseases*.

As this classification is regarded the pioneering attempt to systematize rare cardiovascular diseases the authors of this textbook are, indeed, aware of its imperfections and limitations. Therefore, presenting this classification we sincerely encourage the Readers to provide their solid feedback. Constructive contributions will be recognized.

Conflict of interest: none declared.

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Please note that the RCD Classification is currently under the second round of review by the national and international CRCD experts. For individual author contributions, please see the respective RCD Classes/Sections as per the textbook Parts and Chapters that follow.

Table 1. Classification of Rare Cardiovascular Diseases, Krakow 2013

Group	Subgroup	Examples	RCD code	ICD-10 code
Rare diseases of systemic circulation – class I				
1. Anatomical malformations of the arteries	A. Cerebral arteries	1. Anomalies of the circle of Willis	I-1A.1	Q28.3
		2. Intracerebral arteries	I-1A.2	I67.8
		3. Moyamoya disease	I-1A.3	I67.5
		– Others	I-1A.O	
	B. Aorta and aortic arch main branches	1. Right aortic arch	I-1B.1	Q25.4
		2. Double aortic arch	I-1B.2	Q25.4
		3. Aortic rings	I-1B.3	Q25
		4. Interruption of aortic arch	I-1B.4	Q25.4
		5. Variants in aortic arch arteries	I-1B.5	Q25
		6. Coarctation of the aorta	I-1B.6	Q25.1
		– Others	I-1B.O	
	C. Coronary arteries	1. Variants in the course and the number	I-1C.1	Q24.5
		2. Single coronary artery	I-1C.2	Q24.5
		3. Coronary artery originating from the pulmonary artery	I-1C.3	Q24.5
		4. Coronary fistula	I-1C.4	Q24.5
		5. Coronary artery aneurysm	I-1C.5	Q24.5
		– Others	I-1C.O	
	D. Other arteries	1. Abdominal aorta: cephalic trunk, renal, mesenteric, splenic, others	I-1D.1	Q27.2
		2. Iliac and femoral arteries	I-1D.2	Q27.8
		3. Popliteal and below the knee	I-1D.3	Q27.8
		4. Upper extremity arteries	I-1D.4	Q27.8
		– Others	I-1D.O	
2. Connective tissue disorders causing aneurysmal disease	A. Aneurysmal disease of the aorta	1. Marfan syndrome	I-2A.1	Q87.4
		2. Ehlers–Danlos syndrome	I-2A.2	I71
		3. Loeys–Dietz syndrome	I-2A.3	Q79.6
		4. Familial thoracic aortic aneurysms and dissections	I-2A.4	Q87.4
		- Others	I-2A.O	
	- Others		I-2.O	
3. Autoimmune vascular diseases	A. Primary systemic vasculitis: Predominantly large arteries	1. Takayasu's arteritis	I-3A.1	M31.4
		2. Giant-cell arteritis	I-3A.2	M31.6
		3. Isolated aortitis	I-3A.3	I77.6
		– Others	I-3A.O	

Group	Subgroup	Examples	RCD code	ICD-10 code
3. Autoimmune vascular diseases	A. Primary systemic vasculitis: Predominantly medium-and small-size arteries	5. Kawasaki disease	I-3A.5	M30.3
		6. Polyarteritis nodosa	I-3A.6	M30
		7. Necrotizing ANCA-associated:	I-3A.7	
		a. Churg–Strauss syndrome	I-3A.7a	M31
		b. Wegener's granulomatosis	I-3A.7b	M31.3
		c. Microscopic polyangiitis	I-3A.7c	M31.7
		d. Idiopathic necrotizing crescentic glomerulonephritis	I-3A.7d	N05.7
		8. Non-ANCA associated:	I-3A.8	
		a. Henoch–Schönlein purpura	I-3A.8a	D69
		b. Goodpasture's disease	I-3A.8b	M31.0
		c. Mixed cryoglobulinemia	I-3A.8c	D89.1
		d. Hypersensitivity vasculitis	I-3A.8d	M31.0
		– others	I-3A.8.o	
	B. Secondary systemic vasculitis	1. Secondary to infection (unknown)	I-3B.1	I77.6
		a. Viral	I-3B.1.a	
		b. Bacterial	I-3B.1.b	
		c. Fungal	I-3B.1.c	
		d. Parasitosis	I-3B.1.d	
		2. Secondary to medications	I-3B.2	
	C. Connective tissue disorders causing premature thrombosis / atherosclerosis	1. Systemic lupus erythematosus	I-3C.1	M32
		2. Scleroderma	I-3C.2	M34
		3. Antiphospholipid syndrome	I-3C.3	D68.6
		– Others	I-3C.O	
	– Others	1. Behçet's disease	I-3O.1	M35.2
		2. Cogan syndrome	I-3O.2	Q30.8
		3. Others	I-3O.O	
4. Intimal hyperplasia	A. Fibromuscular dysplasia		I-4A	I77.3
	- Others		I-4O	
5. Spontaneous dissection of the artery	A. Dissection of aortic arch arteries		I-5A	I71.0
	- Others		I-5O	
6. Premature atherosclerosis	A. Familial hypercholesterolemia		I-6A.1	E78
	B. Adult progeria – laminopathies	1. Hutchison–Gilford progeria syndrome	I-6B.1	E34.8
		2. Dunnigan-type partial lipodystrophy	I-6B.2	E88.1
		– Others	I-6B.O	
	C. Secondary	1. Polycystic ovary syndrome	I-6C.1	E28.2
		2. Acquired immunodeficiency syndrome	I-6C.2	B22.2
		– Others	I-6C.O	
– Others			I-O	

Group	Subgroup	Examples	RCD code	ICD-10 code
Rare diseases of pulmonary circulation – RCD class II				
1.Pulmonary hypertension	A. Low-prevalence pulmonary hypertension	1. Idiopathic PAH	II-1A.1	I27
		2. Heritable PAH	II-1A.2	I27
		3. Drug- and toxin-induced PAH	II-1A.3	I27.2
		4. PAH associated with:		
		a. connective tissue disease	II-1A.4a	I27.2
		b. HIV infection	II-1A.4b	I27.2
		c. portal hypertension	II-1A.4c	I27.2
		d. congenital heart diseases	II-1A.4d	I27.2
		– others	II-1A.4.o	
		5. Chronic thromboembolic pulmonary hypertension	II-1A.5	I27.2
		6. Pulmonary veno-occlusive disease	II-1A.6	I27
		7. Pulmonary hemangiomatosis	II-1A.7	D18
		8. Persistent pulmonary hypertension of the newborn	II-1A.8	P29
		– Others	II-1A.O	
	B. Severe forms of non-low-prevalence pulmonary hypertension	1. Severe pulmonary hypertension due to left heart diseases	II-1B.1	I27
		2. Severe pulmonary hypertension due to lung diseases and/or hypoxia	II-1B.2	I27.2
	C. Overlap pulmonary hypertension	1. Pulmonary hypertension in a patient with congenital shunt and left ventricular dysfunction	II-1C.1	I27
		2. Pulmonary hypertension associated with congenital heart disease complicated by thromboembolic disease	II-1C.2	I27
		– Others	II-1C.O	
2. Inborn anomalies of the pulmonary vessels	A. Anomalous morphology	1. Atresia of the pulmonary artery	II-2A.1	Q25.5
		2. Pulmonary artery coarctation	II-2A.2	Q25.7
		3. Idiopathic dilatation of the pulmonary trunk	II-2A.3	Q25
		– Others	II-2A.O	
	B. Anomalous course	1. Pulmonary artery sling	II-2B.1	Q25.6
		2. Ductal sling	II-2B.2	Q33.2
		3. Pulmonary sequestration	II-2B.3	E25.7
		– Others	II-2B.O	
	C. Anomalous connections	1. Inborn pulmonary arteriovenous fistulas	II-2C.1	Q25.7
		– Others	II-2C.O	
3. Acquired anomalies of the pulmonary vessels	A. Pulmonary vessel arteritis	1. Takayasu's arteritis	II-3A.1	M31.4
		2. Giant-cell arteritis	II-3A.2	M31.6
		3. Behçet's disease	II-3A.3	M35.2
		4. Hughes–Stovin syndrome	II-3A.4	M35.2
		5. Granulomatous vasculitis	II-3A.5	M31.3
		– Others	II-3A.O	
	B. Anomalous morphology	1. Pulmonary artery aneurysm	II-3B.1	E25.7
		– Others	II-3B.O	

Group	Subgroup	Examples	RCD code	ICD-10 code
3. Acquired anomalies of the pulmonary vessels	C. Anomalous connections	1. Pulmonary arteriovenous fistulas	II-3C.1	I77
		2. Bronchial artery–pulmonary artery fistulas	II-3C.2	Q27
		– Others	II-3C.O	
	D. Tumors of the pulmonary vessels	1. Primary	II-3D.1	
		2. Secondary	II-3D.2	
		– Others	II-O	

Group	Subgroup	Examples	RCD code	ICD-10 code
Rare diseases of the heart (cardiomyopathies) – RCD class III				
1. Dilated cardiomyopathy	A. Genetic	1. Sarcomeric protein mutations: β-myosin heavy chain (MYH7; on chromosome 14q12), myosin-binding protein C (MYBPC3; 11p11.2), troponin T (TNNT2; 1q32), troponin C (TNNC1; 3p21.3-p14.3), α-myosin heavy chain (MYH6; 14q12), α-tropomyosin (TPM1; 15q22.1), cardiac actin (ACTC; 15q14), and titin (TTN)	III-1A.1	I42.4
		– Other		
		2. Z-band mutations	III-1A.2	I42.4
		3. Cytoskeletal gene mutations:	III-1A.3	I43
		a. Dystrophin – Duchenne muscular dystrophy	III-1A.3a	G71.0
		b. Dystrophin – Becker's muscular dystrophy	III-1A.3b	G71.0
		c. Dystrophin – Bethlem myopathy	III-1A.3c	G71.0
		d. Dystrophin – Limb-girdle muscular dystrophy	III-1A.3d	G71.0
		e. Tafazzin – Barth syndrome	III-1A.3e	E71.1
		f. Desmin mutations	III-1A.3f	G71.8
		g. Sarcoglycan complex mutations	III-1A.3g	G71.0
		– Other cytoskeletal gene mutations	III-1A.3.o	
		4. Nuclear membrane mutations:	III-1A.4	I42.4
		a. Lamins A/C – DCM + conduction disease	III-1A.4a	G71.0
		b. LaminsA/C – Emery–Dreifuss muscular dystrophy	III-1A.4b	G71.0
		– Other nuclear membrane mutations	III-1A.4.o	
		5. Mitochondrial cardiomyopathies	III-1A.5	I43
		a. Kearns–Sayre syndrome	III-1A.5a	H49.8
		– Other mitochondrial cardiomyopathies	III-1A.5.o	
	B. Nongenetic	1. Inflammatory cardiomyopathy:	III-1B.1	I42.7
		a. Viral inflammatory cardiomyopathy	III-1B.1a	B33.24
		b. Nonviral inflammatory cardiomyopathy	III-1B.1b	I42.7
		c. Autoimmune-induced inflammatory cardiomyopathy	III-1B.1c	I42.7
		– Other inflammatory cardiomyopathies	III-1B.1.o	
		2. Due to connective tissue diseases:	III-1B.2	I43
		a. Systemic lupus erythematosus	III-1B.2a	M32
		b. Scleroderma	III-1B.2b	M34
		c. Giant-cell arteritis	III-1B.2c	M31.6
		– Other due to connective tissue diseases	III-1B.2.o	
		3. Due to endocrine disorders:	III-1B.3	I43
		a. Thyroid hormone excess or deficiency	III-1B.3a	E00-07
		b. Pheochromocytoma	III-1B.3b	C75.5/D35.6
		c. Cushing's disease	III-1B.3c	E24
		– Other due to endocrine disorders	III-1B.3.o	
		4. Due to infiltrative disorders:	III-1B.4	I43
		a. Amyloidosis	III-1B.4a	E85
		b. Sarcoidosis	III-1B.4b	D86
		c. Hemochromatosis	III-1B.4c	E83.1
		– Other due to infiltrative disorders	III-1B.4.o	

Group	Subgroup	Examples	RCD code	ICD-10 code
1. Dilated cardio-myopathy	B. Nongenetic	5. Medication-induced:	III-1B.5	I42.7
		a. Anthracyclines	III-1B.5a	I42.7
		b. Cyclophosphamide	III-1B.5b	I42.7
		c. Trastuzumab	III-1B.5c	I42.7
		d. HAART-HIV: zidovudine, didanosine, zalcitabine	III-1B.5d	I42.7
		– Other	III-1B.5.o	
		6. Toxin-induced:	III-1B.6	I42.7
		a. Ethanol	III-1B.6a	I42.6
		b. Cocaine	III-1B.6b	I42.7
		c. Amphetamines	III-1B.6c	I42.7
		– Other	III-1B.6.o	
		7. Tachycardia-induced:	III-1B.7	I42.8
		a. Uncontrolled atrial fibrillation	III-1B.7a	I48
		b. Atrioventricular nodal reentry	III-1B.7b	I47.1
		c. Preexcitation syndromes	III-1B.7c	I45.6
		– Other	III-1B.7.o	
		8. End stage of other types of cardiomyopathy:	III-1B.8	I42.9
		a. Hypertrophic cardiomyopathy	III-1B.8a	I42.2
		b. Restrictive cardiomyopathy	III-1B.8b	I42.5
		c. Peripartum cardiomyopathy	III-1B.8c	O90.3
		d. Takotsubo cardiomyopathy	III-1B.8d	I51.81
		e. Left ventricular noncompaction	III-1B.8e	I42.8
		– Other	III-1B.8.o	
		9. Miscellaneous:	III-1B.9	I43
		a. Neoplastic heart disease	III-1B.9a	D15.1
		b. Celiac disease	III-1B.9b	K90
		c. Extensive chest radiation	III-1B.9c	Y84.2
		d. Nutritional (thiamine, selenium, L-carnitine)	III-1B.9d	I43.2
		e. Obstructive sleep apnea	III-1B.9e	G47.3
		– Other	III-1B.9.o	
2.Hypertrophic cardiomyopathy	A. Sarcomeric protein mutations	1. MYH7, MYBPC3, TNNT2, MYH6, TPM1, TNNC1, ACTC, TTN	III-2A.1	I42.2
	B.Nonsarcomeric protein mutations	1. Glycogen storage disease:	III-2B.1	I43.1
		a. Pompe disease	III-2B.1a	E74.0
		b. Danon disease	III-2B.1b	E74.0
		c. Forbes disease	III-2B.1c	E74.0
		– Other	III-2B.1.o	
		2. Lysosomal storage disease:	III-2B.2	I43.1
		a. Fabry disease	III-2B.2a	E75.2
		b. Hurler syndrome	III-2B.2b	E76
		c. Hunter syndrome	III-2B.2c	E76.1
		d. Maroteaux–Lamy disease	III-2B.2d	E76.2
		e. Gangliosidosis	III-2B.2e	E75.1
		f. Gaucher's diseases	III-2B.2f	E75.2
		g. Niemann–Pick disease	III-2B.2g	E75.2
		– Other	III-2B.2.o	
		3. Metabolic myopathies:	III-2B.3	I43.1
		a. Disorders of fatty metabolism	III-2B.3a	E78
		b. Carnitine deficiency	III-2B.3b	E71.3
		c. Phosphorylase-b kinase deficiency	III-2B.3c	E74
		– Other	III-2B.3.o	
		4. Systemic diseases:	III-2B.4	I43
		a. Pheochromocytoma	III-2B.4a	C75.5/D35.6
		b. Neurofibromatosis	III-2B.4b	Q85.0
		c. Tuberous sclerosis	III-2B.4c	Q85.1
		– Other	III-2B.4.o	
		5. Mitochondrial cardiomyopathies	III-2B.5	I43

Group	Subgroup	Examples	RCD code	ICD-10 code
2. Hypertrophic cardiomyopathy	B. Nonsarcomeric protein mutations	6. Syndromic HCM:	III-2B.6	I43
		a. Noonan syndrome	III-2B.6a	Q87.1
		b. LEOPARD syndrome	III-2B.6b	Q87.8
		c. Friedreich's ataxia	III-2B.6c	G11.1
		d. Swyer syndrome	III-2B.6d	Q97.3
		e. Costello syndrome	III-2B.6e	Q87.8
		– Other	III-2B.6.o	
3. Restrictive cardiomyopathy	A. Infiltrative	1. Familial amyloidosis	III-3A.1	E85
		a. Transthyretin	III-3A.1a	E85.1
		b. Apolipoprotein	III-3A.1b	E85
		2. Amyloid	III-3A.2	E85.1
		a. AL/prealbumin	III-3A.2a	E85.1
		3. Sarcoidosis	III-3A.3	D86
		4. Gaucher's disease	III-3A.4	E75.2
		5. Hurler syndrome	III-3A.5	E76
		6. Fatty infiltration	III-3A.6	E78
		– Other	III-3A.O	
	B. Storage	1. Hemochromatosis	III-3B.1	E83.1
		2. Fabry disease	III-3B.2	E75.2
		3. Glycogen storage disease	III-3B.3	E74
		– Other	III-3B.O	
	C. Noninfiltrative	1. Scleroderma	III-3C.1	M34
		2. Pseudoxanthoma elasticum	III-3C.2	Q82.8
		– Other	III-3C.O	
	D. Sarcomeric protein mutations	Troponin I, essential light chain of myosin	III-3D	I43
	E. Desminopathy		III-3E	G71.8
	F. Endocardial pathology	1. Endomyocardial fibrosis with hypereosinophilia:	III-3F.1	I42.3
		a. Parasitic infection	III-3F.1a	
		b. Drugs – methysergide	III-3F.1b	
		c. Persistent inflammation	III-3F.1c	
		d. Nutritional factors	III-3F.1d	
		2. Endomyocardial disease without hypereosinophilia	III-3F.2	I42.3
		– Other	III-3F.O	
4. Arrhythmogenic right ventricular cardiomyopathy	A. Desmosomal ARVC	1. Autosomal dominant inheritance pattern:	III-4A	I42.8
		a. ARVD8 – Desmoplakin mutations	III-4A.1a	
		b. ARVD9 – Plakophilin-2 mutations	III-4A.1b	
		– Other	III-4A.1.o	
		2. Syndromic ARVC (autosomal recessive)	III-4A.2	I42.8
		a. Naxos disease – Plakoglobin mutations	III-4A.2a	Q87.8
		b. Carvajal syndrome – Desmoplakin mutations	III-4A.2b	I42
		c. Alcalai syndrome	III-4A.2c	I42
		– Other	III-4A.2.o	
	B. Nondesmosomal ARVC	1. ARVD1 – Transforming growth factor mutations	III-4B.1	I42.8
		2. ARVD2 – Cardiac ryanodine receptor mutations	III-4B.2	
		– Other	III-4B.O	

Group	Subgroup	Examples	RCD code	ICD-10 code
5. Unclassified cardiomyopathies	A. Left ventricular noncompaction	1. Genetic causes of LVNC:	III-5A.1	I42.9
		a. Tafazzin mutations	III-5A.1a	
		b. Dystrobrevin mutations	III-5A.1b	
		– Other	III-5A.1.o	
		2. Metabolic disorders/genetic syndromes and LVNC	III-5A.2	I42.9
		a. Barth syndrome	III-5A.2a	E71.1
		b. Beals syndrome	III-5A.2b	Q87.8
		c. Becker's muscular dystrophy	III-5A.2c	G71
		d. Charcot–Marie–Tooth disease	III-5A.2d	G60
		e. Duchenne muscular dystrophy	III-5A.2e	G71
		f. Melnick-needles syndrome	III-5A.2f	Q77.8
		g. Myotonic dystrophy	III-5A.2g	G71.1
		h. Myoadenylate deaminase deficiency	III-5A.2h	E79.8
		i. Nail-patella syndrome	III-5A.2i	Q87.2
		j. Noonan syndrome	III-5A.2j	Q87.1
		k. Roifman syndrome	III-5A.2k	D81.8
		l. Trisomy 13	III-5A.2l	Q90
		– Other	III-5A.2.o	
	B. Takotsubo cardiomyopathy		III-5B	I42.8
	C. Peripartum cardiomyopathy		III-5C	O90.3

Rare congenital cardiovascular diseases – RCD class IV				
1. Abnormalities of the position and connection of the heart and vessels	A. Heart position	1. Dextrocardia	IV-1A.1	Q24.0
		2. Mesocardia	IV-1A.2	Q24.8
		3. Dextroposition	IV-1A.3	Q20.3
		4. Ectopia cordis	IV-1A.4	Q24.8
		– Others	IV-1A.O	
	B. Heart chambers	1. Atria	IV-1B.1	
		a. Cor triatriatum	IV-1B.1a	Q24.2
		– others	IV-1B.1.o	
		2. Ventricles	IV-1B.2	
		a. Congenitally corrected transposition of the great artery	IV-1B.2a	Q20.5
		– others	IV-1B.2o	
	C. Veins and arteries	1. Systemic veins	IV-1C.1	
		a. Left superior vena cava	IV-1C.1a	Q26.1
		– others	IV-1C.1.o	
		2. Pulmonary veins	IV-1C.2	
		a. Pulmonary vein stenosis	IV-1C.2a	Q26.2
		– others	IV-1C.2.o	
		3. Great arteries	IV-1C.3	
		a. Transposition of the great arteries	IV-1C.3a	Q20.3
		b. Truncus arteriosus	IV-1C.3b	
		– others	IV-1C.3.o	Q20.0
	D-Valves	1. Right heart valves	IV-1D.1	
		a. tricuspid atresia	IV-1D.1a	Q22.4
		b. Ebstein's anomaly	IV-1D.1b	Q22.5
		c. pulmonary valve atresia	IV-1D.1c	Q22.0
		d. pulmonary valve stenosis	IV-1D.1d	Q22.1
		– others	IV-1D.1o	

Group	Subgroup	Examples	RCD code	ICD-10 code
1. Abnormalities of the position and connection of the heart and vessels	D-Valves	2. Left heart valves	IV-1D.2	
		a. mitral stenosis	IV-1D.2a	Q23.2
		b. mitral subvalvular apparatus abnormalities	IV-1D.2b	Q23.8
		c. aortic stenosis	IV-1D.2c	Q23.0
		d. aortic regurgitation,	IV-1D.2d	Q23.1
		– others	IV-1D.2o	
2. Shunts	A. Decreased pulmonary flow	1. Tetralogy of Fallot	IV-2A.1	Q21.3
		2. Pulmonary stenosis and ventricular septal defect	IV-2A.2	Q21.3
		3. Pulmonary atresia and ventricular septal defect	IV-2A.3	Q25.5
		– Others	IV-2A.O	
	B. Increased pulmonary flow	1. Atrial septum	IV-2B.1	Q21.1
		2. Atrioventricular junction	IV-2B.2	Q21.2
		3. Ventricular septum	IV-2B.3	Q21.0
		4. Aortopulmonary communication	IV-2B.4	Q21.4
		– Others	IV-2B.O	
3. Complex congenital cardiovascular diseases	A. Complex abnormalities of the position and connection of the heart and vessels		IV-3A	Q20
	B. Complex abnormalities of position and connection of the heart and vessels with shunts		IV-3B	Q20
	– Others		IV-3.O	
4. Congenital cardiovascular diseases with concomitant organ dysfunction	A. Nervous system		IV-4A	G00-99
	B. Pulmonary system		IV-4B	J00-99
	C. Endocrine system		IV-4C	E00-90
	D. Thrombosis and hemostasis disorders		IV-4D	D65-69
	– Others		IV-4.O	
5. Grown-up congenital cardiovascular diseases	A. After correction	1. No complication without residual defects	IV-5A.1	Z92.4
		2. Postprocedural complication and residual defects	IV-5A.2	Z92.4
		– Others	IV-5A.O	
	B. After palliative procedures	1. Fontan procedure	IV-5B.1	Z92.4
		2. Systemic-pulmonary anastomosis	IV-5B.2	Z92.4
		– Others	IV-5B.O	
	C. Uncorrectable		IV-5C	
6. Others		1. Double-chambered left ventricle	IV-6.1	Q20
		– Others	IV-6.O	

Group	Subgroup	Examples	RCD code	ICD-10 code
Rare arrhythmias – RCD class V				
1. Arrhythmias due to primary electrical diseases of the heart	A. Channellopathies	1. Brugada syndrome	V-1A.1	I47.2
		2. Long QT syndrome (LQTS)	V-1A.2	I45.8
		3. Short QT syndrome (SQTS)	V-1A.3	I45.8
		4. Catecholaminergic polymorphic ventricular tachycardia	V-1A.4	I45.8
		– Others	V-1A.0	
	B. Preexcitation syndromes	1. Wolff–Parkinson–White syndrome	V-1B.1	I45.6
		2. Mahaim syndrome	V-1B.2	I45.6
		– Others	V-1B.0	
	– Others		V-10	
2. Arrhythmias secondary to rare structural diseases of the heart	A. In the course of cardiomyopathies	1. Arrhythmogenic right ventricular dysplasia/ cardiomyopathy	V-2A.1	I42.8
		2. Hypertrophic cardiomyopathy	V-2A.2	I42
		3. Restrictive cardiomyopathy	V-2A.3	I42
		4. Left ventricular noncompacted cardiomyopathy	V-2A.4	I42.8
		5. Dilated cardiomyopathy	V-2A.5	I42
		– Others	V-2A.0	
	B. Due to congenital heart diseases	1. Univentricular heart	V-2B.1	Q20.4
		2. Shunts	V-2B.2	
		3. Cor triatriatum	V-2B.3	Q24.2
		4. Persistent left superior vena cava	V-2B.4	Q26.1
		– Others	V-2B.0	
3. Arrhythmias of atypical mechanism and ECG presentation	A. Supraventricular	1. Atypical atrioventricular nodal recurrent tachycardia (AVNRT)	V-3A.1	I47.1
		2. Tachycardia with RP interval longer than PR	V-3A.2	I.47.1
		3. Antidromic atrioventricular tachycardia in Wolff–Parkinson–White syndrome	V-3A.3	I45.6
		4. Tachycardia in Mahaim syndrome	V-3A.4	I.47.1
		– Others	V-3A.0	
	B. Ventricular	1. Bundle branch reentry tachycardia	V-3B.1	I.47.2
		– Others	V-3B.0	
	– Others		V-30	
4. Arrhythmias in rare and specific clinical settings	A. Iatrogenic	1. Cardiotoxicity of chemotherapy	V-4A.1	Z51.1
		2. Post heart transplantation	V-4A.2	Z94.1
		3. Postsurgical correction of congenital heart diseases	V-4A.3	Y83
		- Others	V-4A.0	
	B. Metabolic disorders	1. Fabry disease	V-4B.1	E75.2
		2. Niemann–Pick disease	V-4B.2	E75.2
		– Others	V-4B.0	
	– Others		V-40	

Group	Subgroup	Examples	RCD code	ICD-10 code
Cardiac tumors and cardiovascular diseases in malignancy – RCD class VI				
1. Primary cardiac tumors	A. Primary benign tumors	1. Myxoma	VI-1A.1	D15.1
		2. Fibroma	VI-1A.2	D15.1
		3. Lipoma a. Lipomatous hypertrophy – others	VI-1A.3 VI-1A.3a VI-1A.3.o	D17.0
		4. Rhabdomyoma	VI-1A.4	D21.3
		– Others	VI-1A.O	
	B. Primary malignant tumors	1. Rhabdomyosarcoma	VI-1B.1	C49.3
		2. Angiosarcoma	VI-1B.2	D38.0
		3. Lymphoma	VI-1B.3	C85.9
		4. Hemangioma	VI-1B.4	D18
		– Others	VI-1B.O	
2. Metastatic cardiac tumors	A. Thorax	1. Lung cancer	VI-2A.1	C34.8
		2. Breast cancer	VI-2A.2	C50.8
		– Others	VI-2A.O	
	B. Abdomen	1. Gastrointestinal tract cancer	VI-2B.1	C26.8
		2. Urinary tract and kidney cancer	VI-2B.2	C68.8
		3. Prostate cancer	VI-2B.3	C61
		4. Reproductive system cancer	VI-2B.4	C57.0
		– Others	VI-2B.O	
	C. Hematological system	1. Leukemia	VI-2C.1	C81-96
		2. Lymphoma	VI-2C.2	C81-96
		– Others	VI-2C.O	
	D. Skin cancer		VI-2D	C44
	– Others		VI-2O	
3. Thrombus within heart chambers			VI-3	I74.0
4. Inflammatory malformations	A. Vegetations		VI-4A	I80.9
	B. Inflammatory tumors		VI-4B	R22.6
	C. Abscesses		VI-4C	J85.3
	D. Calcifications	1. Pericardium	VI-4D.1	I32
		2. Valves	VI-4D.2	I39
		– Others	VI-4D.O	
	– Others		VI-4O	
5. Cardiovascular complications of oncological therapy	A. Post-surgery		VI-5A	Y83
	B. Post-radiotherapy		VI-5B	Y84.2
	C. Post-chemotherapy		VI-5C	Z51.1
	– Others		VI-5O	

Cardiovascular diseases in pregnancy – class VII*

rare cardiovascular diseases – main classes (table 1–6)	Following characters for subgroups, examples, according to the tables I–VI and VIII			CRCD code	ICD 10
	Group	Subgroup	Example		
rare diseases of systemic circulation (class I)	1…	A…	1…	VII-I-…	O99.4
rare diseases of pulmonary circulation (class II)	1…	A…	1…	VII-II-…	O99.4
rare diseases of the heart (cardiomyopathies) (class III)	1…	A…	1…	VII-III…	O99.4
rare congenital cardiovascular diseases (class IV)	1…	A…	1…	VII-IV…	O99.4
rare arrhythmias (class V)	1…	A…	1…	VII-V…	O99.4
cardiac tumors and cardiovascular diseases in malignancy (class VI)	1…	A…	1…	VII-VI…	O99.4
Unclassified rare cardiovascular diseases (class VIII)	1…	A…	1…	VII-VIII…	

* The digit VII at the front, indicates class VII and is followed by an appropriate RCD classification code corresponding to a rare cardiovascular entity found in class I to VI. Example: VII-I-1A.1 indicates a pregnant woman with anomaly of the circle of Willis, an entity included in class I of the RCD classification: Class VII – rare cardiovascular diseases in pregnancy, class I – rare diseases of the systemic circulation, group 1 – anatomical malformations of the arteries, subgroup A – cerebral arteries, example 2 – anomalies of the circle of Willis

Unclassified rare cardiovascular diseases – RCD class VIII*

Examples	RCD code
1. 62-year-old woman with Heyde's syndrome.	VIII-1
2. 49-year-old patient with factor VII deficiency, chronic heart failure, and thrombus in the left ventricle.	VIII-2
3. 24-year-old patient with vein thrombosis and thrombus in the apex of the heart during ascariasis.	VIII-3
4. Acute thromboembolic disease complicated with heparin-induced thrombocytopenia type II in a pregnant woman.	VIII-4
5. 47-year-old patient with primary severe tricuspid regurgitation.	VIII-5

* Consecutive unclassified rare cardiovascular cases included in Class VIII are assigned subsequent code according to the order of publication on the CRCD webpage – www.crcd.eu or in the *Journal of Rare Cardiovascular Diseases*.