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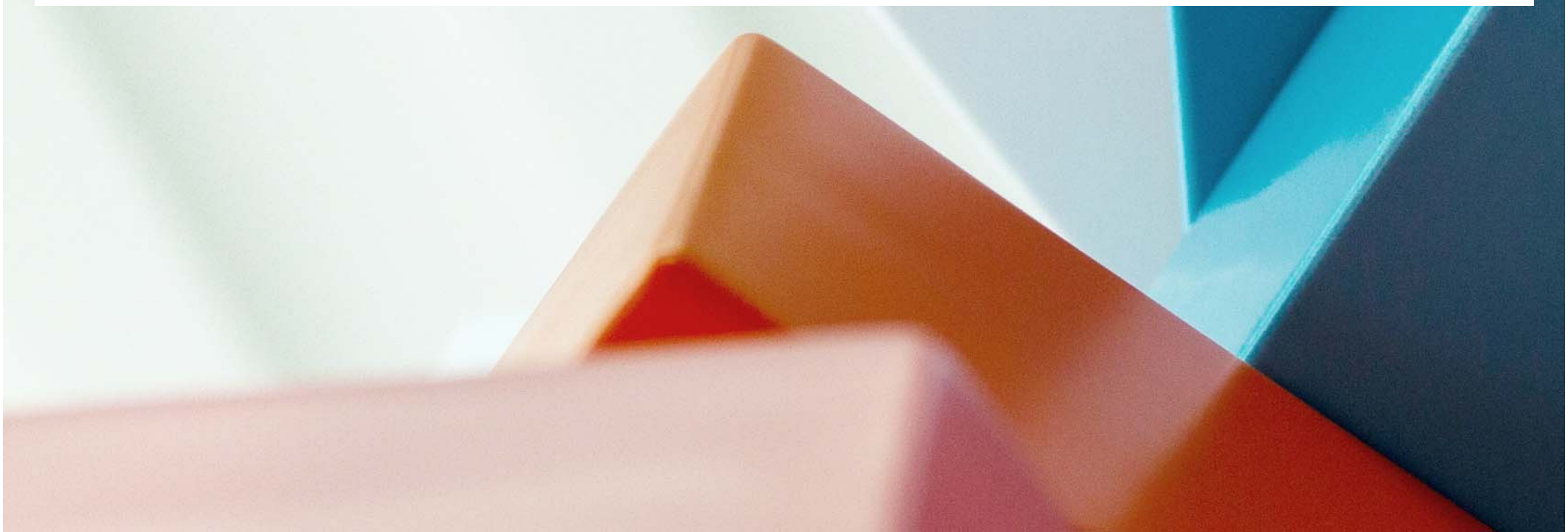
RD terminologiy and coding in Germany

Stefanie Weber, BfArM



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Rare diseases in routine coding

1. ICD-10-GM only has about 350 rare diseases with a specific Code (e.g. Q87.4)

2. Most rare disesas are coded with unspecified codes in ICD-10-GM (e.g. Q87.8)

ICD-10-GM-Tabular list:

Q87.4	Marfan-Syndrom
Q87.5	Sonstige angeborene Fehlbildungssyndrome mit sonstigen Skelettveränderungen
Q87.8	Sonstige näher bezeichnete angeborene Fehlbildungssyndrome, anderenorts nicht klassifiziert
	Alport-Syndrom
	Laurence-Moon-Biedl-Bardet-Syndrom
	Zellweger-Syndrom

Alphabet of ICD-10-GM: 88 Entries for Code Q87.8:

Q87.8	Costello-Syndrom
Q87.8	Dahlberg-Borer-Newcomer-Syndrom
Q87.8	Dahlberg-Syndrom
Q87.8	Desmosterolose
Q87.8	Ellis-Yale-Winter-Syndrom
Q87.8	Fazio-kutano-skelettales Syndrom
Q87.8	Fehlbildungssequenz des urorektalen Septums
Q87.8	German-Syndrom
Q87.8	Hadziselimovic-Syndrom
Q87.8	Hall-Hittner-Syndrom
Q87.8	HEC-Syndrom [Hydrocephalus, endocardial fibroelastosis, cataract]
Q87.8	Hennekam-Syndrom
Q87.8	Hirnfehlbildung mit kongenitalem Herzfehler und postaxialer Polydaktylie
Q87.8	Houlston-Ironton-Temple-Syndrom
Q87.8	Hydrozephalus mit Endokardfibroelastose und Katarakt
Q87.8	Irons-Bianchi-Syndrom

NAMSE Action item 19

NAMSE
Nationales Aktionsbündnis für Menschen mit Seltenen Erkrankungen

Nationaler Aktionsplan
für Menschen mit Seltenen Erkrankungen

Handlungsfelder, Empfehlungen und Maßnahmenvorschläge

Maßnahmenvorschlag 19:

Kodierung aller Patienten mit Seltenen Erkrankungen unter Nutzung des Orpha-Diagnosecodes gekoppelt an ICD-10 GM vor Einführung des ICD-11 im Rahmen eines Projektes zur Implementierung einer einheitlichen Kodierung.

Zeitschiene: kurzfristig
Verantwortliche: Orphanet Deutschland, DIMDI

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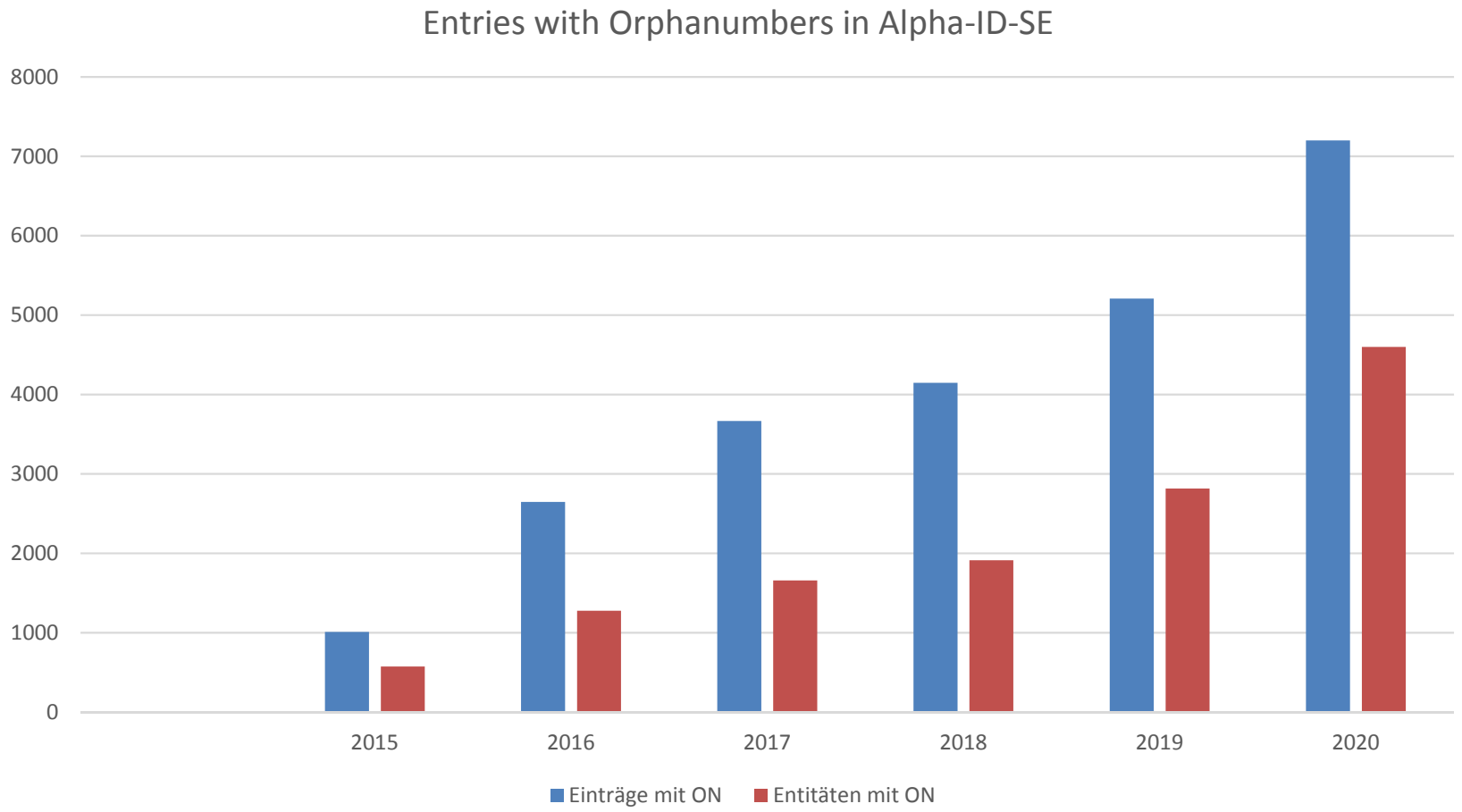
Alpha-ID-SE

Alpha-ID-Kode	ICD-10-GM-Primärkode 1	ICD-10-GM-Stern-Kode	ICD-10-GM-Zusatzkode	ICD-10-GM-Primärkode 2	Orpha-Kennnummer	Eintrag
I4054	L12.1+	H13.3*			99922	Okuläres Pemphigoid
I2461	E78.8+	M14.39*			139436	Lipoiddermatoarthritis
I32050	M61.19				337	Fibrodysplasia ossificans progressiva
I81949	M61.19				337	Münchmeyer-Syndrom
I82889	Q41.1				1201	Apfelschalen-Syndrom
I118177	Q41.1				1201	Jejunalatresie
I17291	Q41.9				1201	Dünndarmatresie
I117676	Q87.1				500	Kardiomyopathische Lentiginose
I9222	Q87.8				912	Zerebro-hepato-renales Syndrom
I125150	C56		C97!	C50.9	145	Hereditäres Brust- und Ovarialkrebssyndrom

Alpha-ID-SE



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Status and next steps

- Since 2020 coding with ICD-10-GM and Orphacodes is mandatory for Centres of Expertise to enable statistical analysis of number of patients with rare diseases treated in centres https://www.g-ba.de/beschluesse/4072/
- In preparation: new legislation to allow for capturing of ICD-10-GM-Codes together with Orphacodes in all hospitals in Germany. This will enable:
  - Statistics on numbers of patients
  - Use of codes in electronic health record
  - …

➢ BfArM will continue to maintain and expand the content of the Alpha-ID-SE and will provide it to the users on a routine basis once a year.

Other news on Classifications and Terminologies from Germany

- BfArM will be National Release Centre for SNOMED CT for Germany starting from January 1st 2021
- LOINC will be further translated and put to more standardized use in Germany
- BfArM will host Orphanet Germany from January 1st 2021

➢BfArM will host a National Competence Centre for medical Terminologies

Thank you very much for your attention!

Contact

Federal Institute for Drugs and Medical Devices
Division P16 – Medical Classifications and Centre for Semantics
Kurt-Georg-Kiesinger-Allee 3
D-53175 Bonn

Contact person
Dr. Stefanie Weber
Stefanie.weber@bfarm.de
www.bfarm.de
Tel. +49 (0)228 99 307-4945

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