



How to improve the codification of RD with ICD-10 and Orphacodes

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Introduction

The estimation of the impact of rare diseases (RD) in Health Care Systems, which is based on their statistical representation, is not satisfying¹. Since there is only few epidemiological data available, numbers for incidence and prevalence of RD are mostly estimated². The prerequisite for an adequate statistical representation of a disease is a consistent and standardised encoding of collected data. This problem affects RD not only in Germany but in all countries around the world.

DIMDI's project „Kodierung von Seltenen Erkrankungen“ (RD coding) is an initiative in the context of the National Action Plan for People with Rare Diseases. It is financed by the Federal Ministry of Health and should improve the statistical data collection and thus the representation of RD in the Health System by mapping the Orphanet Nomenclature and the ICD-10-GM into one file, the **„Musterdatensatz“**. This file should ensure an easy and **standardised** procedure for the daily routine of **double coding** RD with **ICD-10-GM and OrphaNumber** in one step.

To prepare this file, the Orphanet Nomenclature is being mapped to the Alphabetical Index of ICD-10-GM. A dual control principle ensures the quality of this mapping. The process is performed in collaboration with Orphanet. Figure 1 shows an example of the „Musterdatensatz“.

Results and Discussion

The „Musterdatensatz“ is an electronic file that contains a 1:1 mapping of the ICD-10-GM to the OrphaNumber. The content of this file was defined based on the granularity of the Master file of the RD-Action. This file is in ongoing development and currently includes 4.148 entries for RD. The inclusion of all Orphacodes of the Master file is a priority for the further development of the „Musterdatensatz“. The completeness of the file will make the tests more reliable.

The „Musterdatensatz“ was successfully implemented in different ways in the IT systems of four German centres of expertise on RD. In some centres it was possible to integrate it into the hospital coding software.

The centres of expertise reported that the level of granularity of the „Musterdatensatz“ was not adequate for the routine coding of some diseases. That is why it was extended over the level of granularity suggested in the Master file of the RD-Action for some diseases. This is for example the case of the distal myopathies.

The users of the „Musterdatensatz“ are free to use the automatic electronic coding or may use different codes to the proposed on the file. This enables the use of all levels of granularity of both Classifications but risks standardisation and the interoperability of the collected data. An analysis of the collected data and its usability will follow.

Conclusion

- The structure of the „Musterdatensatz“ is mostly satisfying for users.
- The testing and the feedback process of the „Musterdatensatz“ generates valuable input for quality control and further developments of the classifications involved.
- To be able to create a precise connection between the classifications, all involved parties must work together in a coordinated way.
- The Orphanet Nomenclature is updated in a very fast rhythm. This could make the mapping to other classifications difficult and unstable.
- A clear definition of goals, the knowledge of the involved classifications and the selection and preparation of data increases the chance for success.
- The feedback that emerges from the mapping between the classifications as well as from the testing process is a valuable input for further developments and the quality assurance of the classifications.
- The feedback of the coder and the purpose of the process may influence the coding results. This represents a risk for the standardization and thus the interoperability of the collected data.

Methods

The „Musterdatensatz“ is being tested since the middle of 2017 by four centers of expertise on RD which have diverse IT and organizational structures. They implemented the file into their routine coding system and are giving feedback about the usability, structure and content of the file to DIMDI on a regular basis.

The „Musterdatensatz“ can be adapted according to the feedback.

Alpha-ID-Kode	ICD-10-GM Kode	ICD-10-Stern-Kode	ICD-10-Zusatz-Kode	Orpha-Kennnummer	Eintrag
I17291	Q41.9			1201	Dünndarmatresie
I82889	Q41.1			1201	Apfelschalen-Syndrom
I118177	Q41.1			1201	Jejunalatresie
I24937	K90.8+	M14.8*		3452	Whipple-Krankheit
I5821	K90.8			3452	Intestinale Lipodystrophie
I32050	M61.19			337	Fibrodysplasia ossificans progressiva
I81949	M61.19			337	Münchmeyer-Syndrom
I9222	Q87.8			912	Zerebro-hepato-renales Syndrom
I117676	Q87.8			500	Kardiomyopathische Lentiginose

Figure 1: Example of the „Musterdatensatz“

Literatur

[1] CEGRD, 2014. Recommendation on ways to Improve Codification for Rare Diseases in Health Information Systems. Adopted at the 3rd meeting of the Commission Expert Group on Rare Diseases (12.-13. November 2014). [Online] http://ec.europa.eu/health/rare_diseases/docs/recommendation_coding_cegrd_en.pdf [Zugriff am 15 August 2016].

[2] Deutsches Institut für Medizinische Dokumentation und Information (DIMDI); Musterdatensatz, <http://www.dimdi.de/dynamic/de/klassi/downloadcenter/alpha-id/seltene-erkrankungen> Zugriff am 15 August 2016]