A prevalence estimation of exstrophy and epispadias in Germany from public health insurance data

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Background

- Exstrophy-Epispadias-Complex (EEC) is a serious urological birth defect which should be treated in specialized centers.
- Exact prevalence of rare diseases is very important.
- European surveillance of congenital anomalies (EUROCAT) estimates risk for EEC with 1:23,255, i.e. 4.3 per 100,000 births (95%-CI: 3.8 -5.0) during the last decade.
  The range is between 3.0 in 2015 and 6.0 in 2016 (1).
- A Europe-wide prevalence evaluation from excellence centers estimate a prevalence of 3.1 for exstrophy (ICD-10: Q64.1) and of 1,0 for epipadias (ICD-10: Q64.0) (2).
- The German EUROCAT registry in Saxony-Anhalt (covers ~2% of all German births) reports for exstrophy 3.4 for (95%-CI: 1.3 - 6.9) and 2.9 (95%-CI: 1.0 - 6.3) for epispadias(3); i.e. 6.3 for EEC more often the Europe-wide.
- Old estimates are 3.3 for exstrophy and 2.4 for epispadias (4).
- In contrast to these, an insurance reports a decrease for exstrophy from 2.4 to 1.6 and an increase for epispadias from 8.0 to 11.6 (5).
- Prevalence is not necessary an attending prevalence in later years; latter is relevant for after care.

Aims

Exact frequency of exstrophy (ICD-10: Q64.1) and epispadias (ICD-10: Q64.0) treated in different age groups in Germany

Methods

- Each patient is categorized according ICD-10 in German health insurance system.
- These data are now available for research in accordance with §§303a to 303f of the Social Security Code, book five (SGB V) and Data Transparency Ordiance.
- Data are provide by the DIMDI institute as part of the Federal Ministry of Health (BMG) in aggregated manner.
- Benefits:
  - validated by the insurances to protect false bills
  - that they clearly refer to specific individuals
  - quite complete and hence representative for about 71 million, i.e. approx. 87% of the German population.
- There are strong data safety regulations, which limits possible strata or cell size, especially in rare diseases. Hence not all age groups or both gender are possible.
- We reqested the ICD-10 diagnoses for EEC for 2009-2011.

Conclusions

Data are quite robust over the consecutive years 2009-2011. Live time prevalence, especially of epispadias is higher then assumed. Probably milder episapdias are insufficiently recorded in current registries. The current registries seem to underestimate the prevalence due to age related attendance.

Results

The delivered rate for three age groups are presented in the three tables. In Figure 1 we present the calculated prevalence.

Table 1: Total number of EEC adults according to DIMDI

name	year: 2009	year: 2010	year: 2011	Average 2009-2011
18-99 years denominator	59 258 500	59 296 294	59 467 858	59 340 884
Epispadias (Q64.0)	351	363	371	362
Ekstrophy (Q64.1)	747	746	736	743

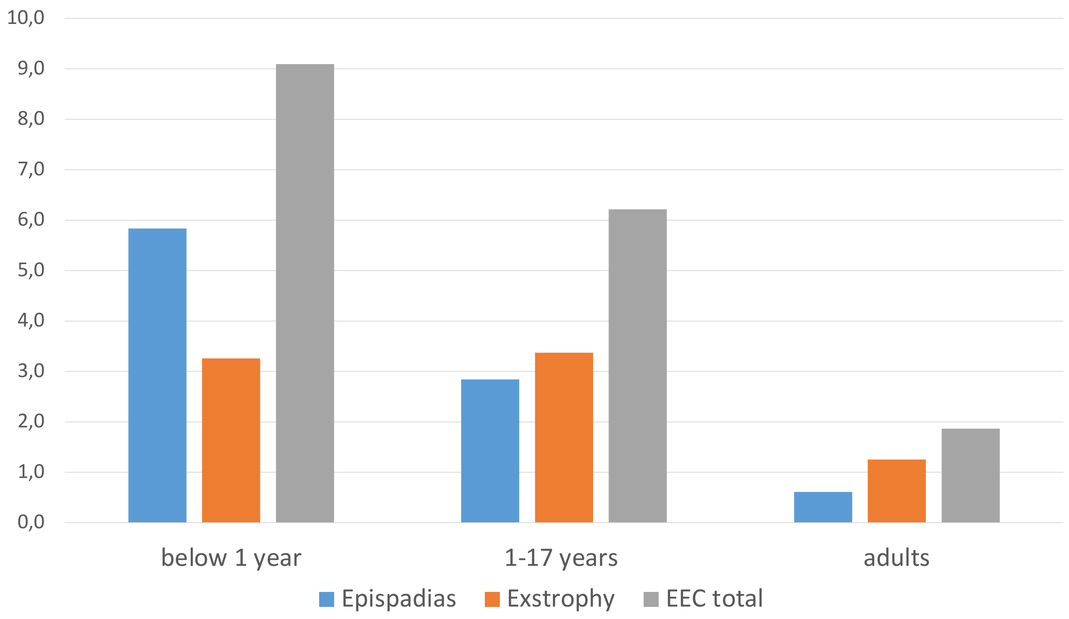
Table 2: Total number of EEC children and adolescents 1-17 years of age according to DIMDI

name	year: 2009	year: 2010	year: 2011	Average 2009-2011
1-17 years denominator	11 226 731	11 070 155	10 975 021	11 090 636
Epispadias (Q64.0)	313	314	318	315
Ekstrophy (Q64.1)	378	379	365	374

Table 3: Newborns eacht year according to DIMDI (precise for prevalence evaluation to surgery need)

name	year: 2009	year: 2010	year: 2011	Average 2009-2011
Babies <1 year	665 126	677 947	662 685	668 586
Epispadias (Q64.0)	35	37	30	34
Ekstrophy (Q64.1)	22	19	15	19

Figure 1: Live Prevalence per 100,000 births according DIMDI data



1. European network of population-based registries for the epidemiological surveillance of congenital anomalies (EUROCAT). 2020. Accessed December 20, 2020: https://eu-rd-platform.jrc.ec.europa.eu/eurocat/eurocat-data/prevalence_en
2. Cervellione RM, Mantovani A, Gearhart J, Bogaert G, Gobet R, Caione P, et al. Prospective study on the incidence of bladder/cloacal exstrophy and epispadias in Europe. J Pediatr Urol. (2015) 11(6):337.e1-6. doi:10.1016/j.jpurol.2015.03.023
3. Götz D, Köhn A, Rißmann A, Spillner C, Vogt C (2020) Jahresbericht des Bundeslandes Sachsen-Anhalt zur Häufigkeit von congenitalen Fehlbildungen und Anomalien sowie genetisch bedingten Erkrankungen 2019. Accessed December 29, 2020: http://www.angeborene-fehlbildungen.com/monz_mm/Dokumente/Jahresberichte/Bericht2019_WEB.pdf
4. Epidemiology of bladder exstrophy and epispadias: A communication from the international clearinghouse for birth defects monitoring systems. Teratology. 1987. doi:10.1002/tera.1420360210
5. Lloyd JC, Wiener JS, Gargollo PC, Inman BA, Ross SS, Routh JC. Contemporary epidemiological trends in complex congenital genitourinary anomalies. J Urol. (2013) 190(4 Suppl):1590-95. doi: 10.1016/j.juro.2013.04.034.