Congenital Anomalies Cork & Kerry

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In this Issue:

References available on request

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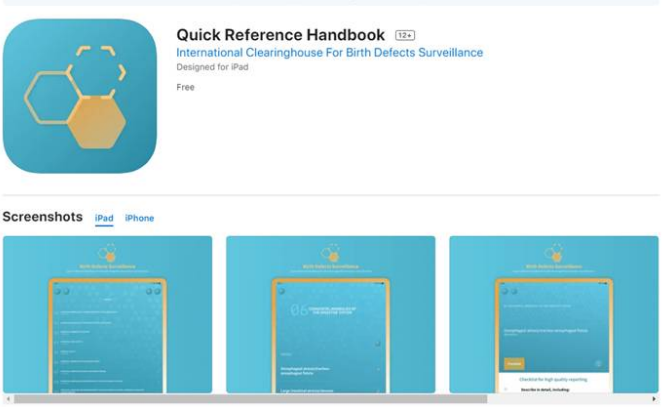
The App of the Quick Reference Handbook (QRH) is out !

An APP for the *Birth defects surveillance: quick reference handbook of selected congenital anomalies and infections,* is now available for both Apple (IOS) and Android systems



The content closely mirrors the original QRH but the app format now allows you to take advantage of typical app features (search, bookmark, tap and enlarge, etc.) – and you can use the QRH anywhere and anytime, when seeing the baby, reviewing records for abstraction, or just when reviewing your knowledge or learning new skills. **Now, checklists, definitions, codes, diagnostic pearls, photos and diagrams are literally at your fingertips.**

To find the app, go to the Apple or Android stores and search for 'QRH birth defects' – for example:

IOS: https://apps.apple.com/ie/app/quick-reference-handbook/id1592730552

Android: https://play.google.com/store/apps/details?id=com.icbdsr.qrh&hl=en_US&gl=US

Multilevel analyses of related Public Health indicators: The European Surveillance of Congenital Anomalies (EUROCAT) Public Health Indicators

Check our publication in the Journal of Paediatric and Perinatal Epidemiology available at following link

https://www.lenus.ie/handle/10147/630081

Data on 55,363 cases with congenital anomalies notified by 18 participating full member EUROCAT Registries in 10 countries between 1st January 2008 and 31st December 2012 were included in this study. A total of 2,430,440 total births during the study period. The following indicators for the 18 registries were pooled according to country.

1. The prevalence of congenital anomaly-related perinatal mortality per 1000 total births
2. The prevalence of termination of pregnancy for foetal anomaly (TOPFA) per 1000 total births
3. The prevalence of prenatal diagnosis of a congenital anomaly per 1000 total births

The study addressed the following question—How can we analyse inter-linked public health indicators?

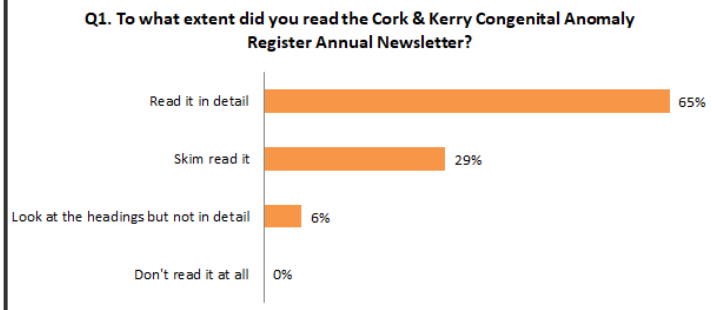
Using multilevel analysis, it is demonstrated that TOPFA prevalence was negatively associated with congenital anomaly-related perinatal mortality prevalence and accounted for 75.5% of between-country variation. Prenatal diagnosis with TOPFA and prenatal diagnosis when modelled together accounted for 83% of between country variation in perinatal mortality. It's important to understand variation in TOPFA rates when interpreting and comparing congenital anomaly-related perinatal mortality rates between countries.

Cork & Kerry Congenital Anomaly Register - Newsletter Evaluation Feedback

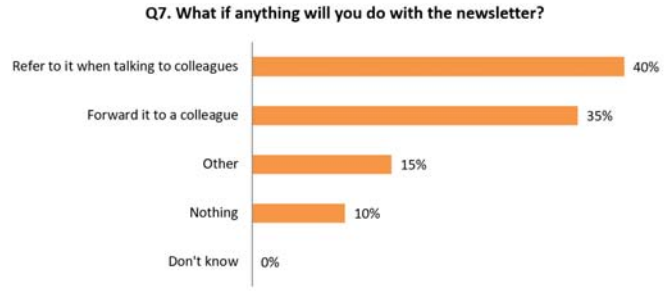
A total of 89 postal questionnaires were circulated to relevant medical staff across Cork/Kerry Acute settings in June 2021. A total of 17 questionnaires were returned giving a response rate of 19%. Of these 53% (n=9) were Consultants, 35% (n=6) Nurses/Midwifes and there were 2 others (12%). For 70% of respondents, the content of the newsletters was considered relevant to them. Factors that may have impacted on the low response rate uptake could be attributed to the cyber-attack and the fact that we had to resort to a postal questionnaire instead of the preferred electronic option. The registry has produced ten Newsletter publication covering the following years (1996-2000, 2006, 2007, 2010, 2014, 2015, 2016, 2017, 2018 and 2020). Follow link below to access all publications. This is the first evaluation of the newsletter conducted by the registry. Survey respondents were circulated with a printed copy of the most recent publication to read - **Congenital Anomaly Register Newsletter Cork & Kerry Newsletter; Vol 9 (1), 2020**, a two-page questionnaire to complete and a stamped addressed envelope to return completed surveys.

A total of 12 questions were asked with a mix of open and tick-box options. The purpose of the survey was to seek feedback from readers of the newsletter on its usefulness and relevance as well as feedback on the aesthetics – look and format. We were also interested in hearing if there were any improvements we could make to the Newsletter content and to promote access to the registry data for further research purposes.
https://www.hse.ie/eng/services/list/5/publichealth/publichealthdepts/howweimprovehealth/congenital-anomaly-registers.html

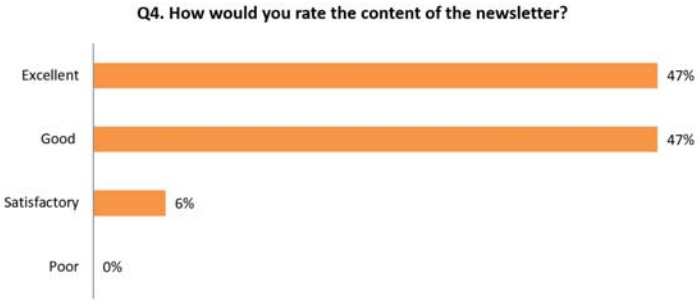
To what extent did you read the Cork & Kerry Congenital Anomaly Register Annual Newsletter?



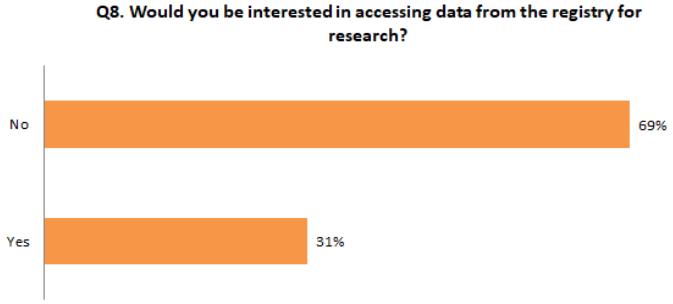
What if anything will you do with the newsletter?



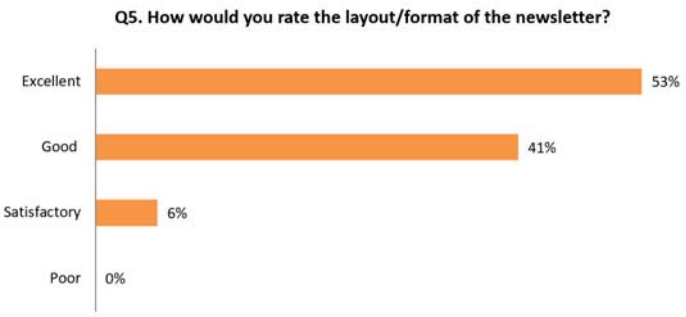
How would you rate the content of the newsletter?



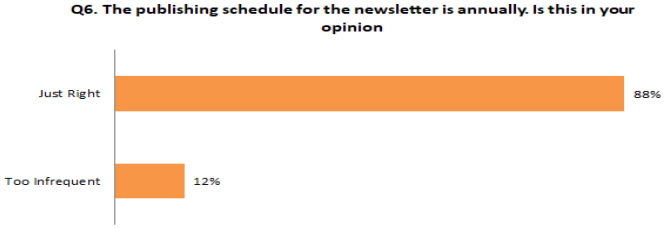
Would you be interested in accessing data from the registry for research?



How would you rate the layout/format of the newsletter?



Is the publishing schedule sufficient?



How could we improve our newsletter?

What information would you like to see included/ removed from the newsletter in the future and other comments.

- Email it out when next edition is available so we can forward it on to staff. Very interesting
- Very informative. No need to change. I think its perfect.
- Excellent very informative newsletter
- Perhaps researchers accessing this data could present their findings

Mortality from Congenital Zika Syndrome — Nationwide Cohort Study in Brazil & Key ZIKA VIRUS DISEASE FACTS from the ECDC 2019 Surveillance Report

The NEW ENGLAND JOURNAL of MEDICINE

Mortality from Congenital Zika Syndrome— Nationwide Cohort Study in Brazil

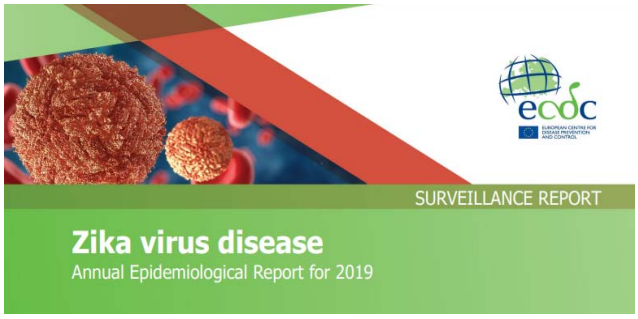
Paixao ES, Cardim LL, Costa MCN, Brickley EB, de Carvalho-Sauer RCO, Carmo EH, et al.

N Engl J Med [Internet]. 2022 February 23;386(8):757–67. Available from: https://doi.org/10.1056/NEJMoa2101195

Methodology: A retrospective population-based cohort study of all singleton live births in Brazil from January 1, 2015, to December 31, 2018. These live-born children were followed from birth until 36 months (3 years), death, or December 31, 2018, whichever occurred first.

Results: A total of 11,481,215 live-born children were followed to 36 months of age. The mortality rate was 52.6 deaths (95% confidence interval [CI], 47.6 to 58.0) per 1000 person-years among live-born children with congenital Zika syndrome, as compared with 5.6 deaths (95% CI, 5.6 to 5.7) per 1000 person-years among those without the syndrome. The mortality rate ratio among live-born children with congenital Zika syndrome, as compared with those without the syndrome, was 11.3 (95% CI, 10.2 to 12.4). Among infants born before 32 weeks of gestation or with a birth weight of less than 1500 g, the risks of death were similar regardless of congenital Zika syndrome status. Among infants born at term, those with con- genital Zika syndrome were 14.3 times (95% CI, 12.4 to 16.4) as likely to die as those without the syndrome (mortality rate, 38.4 vs. 2.7 deaths per 1000 person-years). Among infants with a birth weight of 2500 g or greater, those with congenital Zika syndrome were 12.9 times (95% CI, 10.9 to 15.3) as likely to die as those without the syndrome (mortality rate, 32.6 vs. 2.5 deaths per 1000 person-years). The burden of congenital anomalies, diseases of the nervous system, and infectious diseases as recorded causes of deaths was higher among live-born children with congenital Zika syndrome than among those without the syndrome.

Conclusion: The risk of death was higher among live-born children with congenital Zika syndrome than among those without the syndrome and persisted throughout the first three years of life.



European Centre for Disease Prevention and Control Zika Virus (ZIKV) Disease

https://www.ecdc.europa.eu/sites/default/files/documents/AER-Zika-2019.pdf

Key facts from the EU/EEA countries Annual Epidemiological Report for 2019;

- 71 cases of ZIKV disease were reported in twelve Member States.
- The notification rate for 2019 was 0.02 cases per 100,000.
- In comparison to 0.16 per 100,000 (2018), 0.1 per 100 000 (2017) and 0.6 per 100,000 (2016).
- Five cases of ZIKV disease were reported to have been locally acquired, three autochthonous vector-borne cases, one case of sexual transmission, and one case of non-mosquito borne transmission.
- 92% of cases of ZIKV disease with known importation status were from returning travellers.
- The pregnancy outcome was known for four cases, one of which was born with microcephaly.

The WHO recommends;

Comprehensive information concerning the risk associated with ZIKV infection should be provided before travel.

Travellers to areas with the on-going or historical transmission of ZIKV should take precautions to prevent mosquito bites and avoid/delay pregnancy.

Pregnant women avoid travel to areas with Zika virus transmission based on the increased risk of microcephaly and other severe congenital malformations.

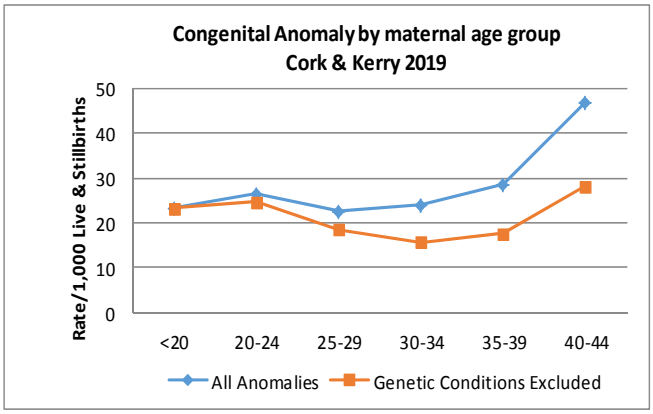
EPIDEMIOLOGY: CORK & KERRY CONGENITAL ANOMALY REGISTER 2019 DATA

There were a total of 8,069 live births (4,115 Males and 3,954 Females) in Cork & Kerry in 2019. There were 34 stillbirths in 2019. The number of babies born with a birth defect was 220 (3%), 18 twin deliveries, 201 singleton and 1 singleton at delivery, but multiple in earlier stage. Of these, there were 118 male, 99 female and 3 babies where the sex was not documented.
Mother's age at delivery where the child had a congenital anomaly in 2019 is shown in Table 1. The prevalence of delivering a baby with a congenital anomaly increases with maternal age from age 35+.

TABLE 1: BIRTHS BY MATERNAL AGE 2019 IN CORK & KERRY

Age Group	All Live Births 2019	Births with Congenital anomalies*	Rate/1000 Live births
15-19	86	2	23.25
20-24	528	14	26.51
25-29	1237	28	22.63
30-34	2748	66	24.01
35-39	2773	79	28.48
40-44	640	30	46.87
45+	57	1	17.54
Total	8069	220	27.26

** 2019 Data extracted from Registry 07th March 2022*



In 9 (26%) of the stillbirths registered in Cork & Kerry during 2019, a congenital anomaly was present. Stillbirths accounted for 0.4% of total births in Cork & Kerry in 2019. There were 29 terminations of pregnancy and 2 spontaneous abortions following prenatal diagnosis.** There were 180 live births of babies with a congenital anomaly in the Cork & Kerry region in 2019. See Table 2 below.

The diagnosis of a congenital anomaly in a child can be identified at birth but in many instances it can be a delayed diagnosis. In 2019, 2 babies with Congenital Anomaly were discovered >12 months.

See Table 3 below when a congenital anomaly diagnosis was discovered in Cork and Kerry babies in 2019.

TABLE 2: TYPE OF BIRTH 2019 CORK & KERRY

Type of Birth	Number of Cases	%
Live birth	180	82%
Stillbirth or fetal death >=20 weeks gestation	9	4%
Termination of Pregnancy	29	13%
Spontaneous Abortion	2	1%
Total number of infants	220	100%

TABLE 3: WHEN DIAGNOSIS WAS DISCOVERED IN CORK & KERRY IN 2019

When Discovered	Cases	%
Prenatal diagnosis	102	46.36%
At birth	71	32.27%
Less than 1 week	25	11.36%
1-4 weeks	5	2.27%
1-12 months	15	6.82%
Over 12 months	2	0.19%
Total	220	100%

In 2019, the gestational age range for babies born with a congenital anomaly ranged between 13 to 42 weeks. A baby born with a congenital anomaly is more likely to be born prematurely and to have a low birth weight. See Tables 4 and 5.

TABLE 4 GESTATIONAL AGE OF INFANTS ON CORK & KERRY REGISTER 2019

Length of Gestation 2019	No (%) of infants registered on Cork & Kerry Register 2019	No (%) of infants nationally 2019
Under 35	58 (26%)	2, 342 (4%)
36 & over	162 (74%)	56, 749 (96%)
Not Stated	0 (0%)	203 (0%)
Total	220 (100%)	59, 294 (100%)

¬ CSO Annual Report on Vital Statistics 2019.
^ Stillbirth data - Not all Stillbirths are registered. Registration of a stillbirth is voluntary .**The numbers recorded by the CSO for Cork & Kerry in 2019 is 16. This is an underestimate of the true number of 34 for this region . As registration is voluntary, only the numbers of stillborn births that are registered are published. This number has been obtained by the Registry from the Birth Notification Form and the Labour Ward Registry .**
**** Termination of Pregnancy for Fetal Anomaly.**

TABLE 5 INFANT BIRTH WEIGHT CORK & KERRY REGISTER 2019

Birth weight 2019	No (%) Infants in Cork & Kerry Register 2019	No (%) of infants nationally 2019¬
Under 2499g	52 (24%)	3289 (6%)
2500g & over	168 (76%)	55834 (94%)
Not stated	0 (0%)	171 (0%)
Total	220 (100%)	59,294 (100%)

See tables 6 and 7 for prevalence and aetiology of congenital anomalies registered for 2019 in our region.

TABLE 6 CASES AND PREVALENCE PER 10,000 BIRTHS FROM CORK & KERRY REGISTRY DATA COMPARED TO EUROCAT FULL MEMBER REGISTRY DATA, 2019

(INCLUDES LIVE BIRTHS, GENETIC CONDITIONS, FETAL DEATHS, AND TERMINATIONS OF PREGNANCY FOR FETAL ANOMALY FOLLOWING PRENATAL DIAGNOSIS WHERE DATA IS AVAILABLE)

Congenital Anomaly	Cork & Kerry Cases	Cork & Kerry Prevalence RATE (95% CI)	EUROCAT Prevalence RATE (95% CI)
ALL ANOMALIES	220	267.86 (233.58 - 305.76)	255.15 (251.10 - 259.24)
Anomaly			
Nervous system	26	31.80 (20.77 - 46.60)	26.95 (25.64 - 28.30)
Eye	6	7.34 (2.66 - 16.01)	3.47 (3.01 - 3.97)
Ear, face and neck	1	1.22 (0.00 - 6.93)	1.38 (1.10 - 1.71)
Congenital heart defects	88	107.63 (86.34 - 132.59)	80.29 (78.02 - 82.60)
Respiratory	7	8.56 (3.41 - 17.67)	4.41 (3.89 - 4.98)
Oro-facial clefts	13	15.90 (8.45 - 27.21)	14.21 (13.26 - 15.20)
Digestive system	13	15.90 (8.45 - 27.21)	18.36 (17.29 - 19.49)
Abdominal wall defects	10	12.23 (5.84 - 22.52)	8.10 (7.39 - 8.85)
Urinary	23	28.13 (17.83 - 42.21)	35.41 (33.92 - 36.96)
Genital	16	19.57 (11.17 - 31.79)	16.98 (15.95 - 18.06)
Limb	52	63.60 (47.51 - 83.40)	39.07 (37.49 - 40.69)
Chromosomal	47	57.49 (42.25 - 76.44)	47.73 (45.99 - 49.52)

*NOTE: Table extracted from the EUROCAT website data prevalence reports on March 7th 2022. The total number of cases on the Cork & Kerry Congenital Anomaly Register are for the year 2019 (n=220) .

TABLE 7 MULTIPLE MALFORMATION OF CONGENITAL ANOMALY 2019 CORK & KERRY

Multiple Malformations 2019	No. Cases	%
Isolated cardiac	44	20%
Genetic syndrome, skeletal dysplasia and monogenic disorder	18	8%
Chromosomal	47	21%
Isolated other	67	31%
Multiple anomalies	26	12%
NTD isolated	4	2%
Isolated renal	11	5%
Teratogenic syndromes (CMV)	3	1%
Total	220	100%

Half of all anomalies are isolated (cardiac 20%; other isolated 31%). Multiple anomalies are present in about one in eight cases (12%). There is a known chromosomal (21%) or other genetic cause (8%) in about 30% of cases. In 2019 there were 3 cases of birth defect secondary to maternal cytomegalovirus (CMV) infection during pregnancy.

A word of thanks to the staff of Cork & Kerry Hospitals who facilitate our Research Nurse in assisting with case ascertainment and accessing medical charts to update the registry.

For further information on Congenital Anomaly Registers in Ireland go to our website at

http://www.hse.ie/congenitalanomalyregistersireland

http://www.eurocat-network.eu/

¬ CSO Annual Report on Vital Statistics 2019