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Health Service Executive

Congenital Anomalies Cork & Kerry

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In this Issue:

References available on request

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EUROCAT Website Working Group Update

The EUROCAT Website Working Group met at JRC ISPRA on 31st August and is happy to announce that they will be launching a revamped and updated website.
http://www.eurocat-network.eu/

eurocat
european surveillance of congenital anomalies

National Maternity System MN-CMS & Cork & Kerry Congenital Anomaly Register

The Maternity & Newborn - Clinical Management System (MN-CMS) has gone live in Cork & Kerry providing an Electronic Health Record (EHR) for all women and babies in maternity services. Prof. Richard Greene, Director of the National Perinatal Epidemiology Centre and UCC's Professor of Clinical Obstetrics, is a member of the MN-CMS project board.

The Cork & Kerry Congenital Anomaly Register (C&KCAR) will be facilitated with read-only access to the new system to facilitate data collection for the registry research nurse. An active approach has been used for data collection since 1996 whereby the research nurse examines data from multiple sources (examination of maternal, new-born, neonatal and paediatric medical charts) to identify cases, confirm the diagnosis and complete the collection of data on core and additional data variables. http://www.hse.ie/congenitalanomalyregistersireland

Maternal & Newborn Clinical Management System
MN-CMS

The C&KCAR Research Nurse is due to receive training on the MN-CMS in the coming months. Access to the electronic maternity system is likely to greatly benefit the register and enhance productivity for the register in the future. The national rollout of the electronic maternity and new-born record could potentially assist the national extension of congenital anomaly surveillance.

Ask About Alcohol & FASD

ask about alcohol.ie

http://www.askaboutalcohol.ie/ was recently launched by the HSE to improve peoples knowledge about alcohol and drinking.
There is a section of the site specific to alcohol and pregnancy with information for parents on Fetal Alcohol Spectrum Disorder (FASD) and Fetal Alcohol Syndrome (FAS) :
http://www.askaboutalcohol.ie/health/alcohol-and-pregnancy/alcohol-and-pregnancy.html

You may wish to check out the following article published online in the Lancet Jan 2017
Estimation of national, regional, and global prevalence of alcohol use during pregnancy and fetal alcohol syndrome: a systematic review and meta-analysis, Popova, Svetlana et al.
The Lancet Global Health , Volume 5 , Issue 3 , e290 - e299
http://www.thelancet.com/journals/langlo/article/PIIS2214-109X%2817%2930021-9/abstract

Zika Surveillance Update

Over the last year, the world has been experiencing an extensive epidemic of Zika virus infection. Since 2007 and as of 5 January 2017, 75 countries and territories have reported evidence of mosquito-borne transmission of Zika virus. Mother to child transmission of Zika virus infection has been definitively linked to an increased likelihood of development of foetal microcephaly, a condition marked by unusually small heads that can result in developmental problems, especially if contracted during the first trimester of pregnancy.

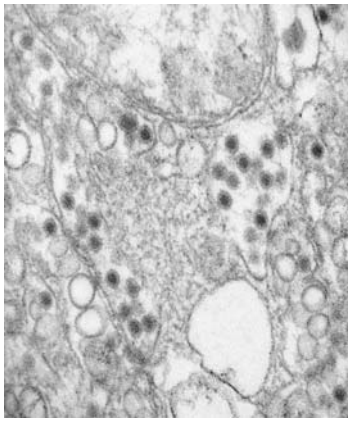


Photo Source: CDC/ National Center for Emerging and Zoonotic Infectious Diseases (NCEZID) Public Health Image Library

The association between Zika virus infection and Guillain-Barré syndrome is also confirmed. The current catastrophic outcome in South America results from the introduction of the infection into an infection naïve population. This contrasts with the situation in Africa, where the infection has been endemic for decades, infection is usually acquired when young with resultant Immunity prior to reproductive age. Women who are pregnant or planning pregnancy are advised to avoid unnecessary travel to affected areas.

Zika is a viral infection that usually causes a mild illness that typically lasts between 2 to 7 days. 80% of people who become infected by Zika virus have no symptoms. The incubation period is 3-12 days. Blood plasma or serum, or urine can be tested within 14 days of first symptoms for viral RNA. Full recovery from the infection is usual. Zika virus infection is primarily spread through the bite of an infected mosquito of *Aedes* genus that is largely tropical and subtropical in distribution. The temperature in Ireland is not conducive to breeding of the Aedes mosquito which needs warmer temperatures.

Zika may be transmitted by sexual or blood exposure to an infected person during the viraemic stage, when the virus is circulating in the blood. Zika virus infection can persist for some time in semen. In order to prevent onward transmission of infection, condom use is recommended for returning male travellers for 6 months following ZIKA infection or for one month after return from an infected area (without symptoms or signs of infection), and for the duration of pregnancy in a partner in order to protect the foetus. Patients with Zika infection do not pose a risk to carers or to healthcare staff. Zika virus infection does not naturally occur in Ireland. However, as of 16th December 2016, there have been a total of 16 cases of Zika infection in persons who have travelled to an affected areas and been diagnosed in Ireland.

On February 1st 2016, World Health Organization (WHO) declared the increase in Zika virus infection and attendant microcephaly to be a Public Health Emergency of International Concern (PHEIC). Zika virus expanded across the Americas, the Pacific Rim and the Caribbean during 2016. Following extensive investigation and the application of widespread control measures in affected countries, WHO declared, on November 18th 2016, the global Zika situation no longer constituted a PHEIC.

The most recent Situational Report from WHO concludes that "Overall, the global risk assessment has not changed. Zika virus continues to spread geographically to areas where competent vectors are present. Although a decline in cases of Zika infection has been reported in some countries, or in some parts of countries, vigilance needs to remain high".

A paper on the prevalence of microcephaly in Europe was recently published in the British Medical Journal by Joan Morris et al. http://www.bmj.com/content/bmj/354/bmj.i4721.full.pdf

HPSC maintains an updated list of countries reporting local transmission of confirmed Zika virus infection. Countries and territories are categorised as currently experiencing active local Zika virus transmission if local Zika infections have been reported by health authorities within the last three months.

Sources: www.hpsc.ie, http://www.who.int/emergencies/zika-virus/situation-report/05-january-2017/en/ http://www.hpsc.ie/A-Z/Vectorborne/Zika/Factsheet/Listofaffectedcountries/

European surveillance of Zika virus in pregnancy

ECDC is exploring enhanced surveillance for ZIKV cases diagnosed in pregnancy based on existing reporting structures (TESSy) in collaboration with national public health institutes (HPSC in Ireland). EUROCAT will collaborate with ECDC by providing information on the epidemiology of microcephaly and other CNS malformations.

EPIDEMIOLOGY: CORK & KERRY CONGENITAL ANOMALY REGISTER 2013 DATA

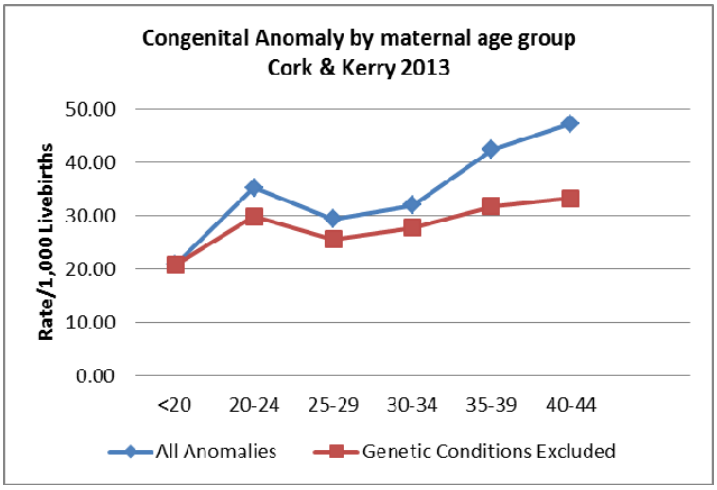
There were 9565 live births (4894 males; 4671 females) in Cork & Kerry in 2013 and 24 stillbirths. The number of babies born with a birth defect was 340 (3.5 %), 316 singleton and 24 twin deliveries. Of these, there were 124 male, 212 female and 4 babies where the sex was indeterminate.

Mother's age at delivery where the child had a congenital anomaly in 2013 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 2013 IN CORK & KERRY

Age Group	All Live Births	Births with Con-genital anoma-lies	Rate/1000 Livebirths
<20	144	3	20.83
20-24	736	26	35.32
25-29	1800	53	29.44
30-34	3563	114	31.99
35-39	2732	116	42.45
40-44	571	27	47.28
45+	19	0	0
Total	**9565**	**339***	**35.44**

* There was one birth with a congenital anomaly where maternal age is not recorded.
Data table extracted from Registry Nov 2016



In 6 (25%) of the stillbirths registered in Cork & Kerry in 2013, a congenital anomaly was present. Stillbirths accounted for 0.25% of total births in 2013. There were 7 terminations of pregnancy following prenatal diagnosis.* There were 327 live births of babies with a congenital anomaly in the Cork & Kerry region in 2013. See Table 2.

TABLE 2: TYPE OF BIRTH 2013 CORK & KERRY

Type of Birth	Number of Cases	%
Live birth	327	96.20%
Stillbirth >=20 weeks gestation	6	1.80%
Termination of Pregnancy	7	2.10%
Total number of infants	**340**	**100%**

The diagnosis of a congenital anomaly in a child can be identified at birth but in many instances it can be a delayed diagnosis. See Table 3 when a congenital anomaly diagnosis was discovered in Cork and Kerry babies in 2013.

TABLE 3: WHEN DIAGNOSIS WAS DISCOVERED 2013 IN

When Discovered	Cases	%
Prenatal diagnosis	66	19.40%
At birth	119	35.00%
Less than 1 week	17	5.00%
1-4 weeks	15	4.40%
1-12 months	119	35.00%
Over 12 months	4	1.20%
Total	**340**	**100%**

In the 2013 the gestational age range for babies born with a congenital anomaly ranged between 21 to 42 weeks. A baby 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 2013

Length of Gestation	No (%) of infants registered on Cork & Kerry Register	No (%) of infants nationally*
Under 35	39 (11%)	2707 (4%)
36 & over	289 (85%)	64266 (93%)
Not Stated	12 (4%)	1981 (3%)
Total	**340 (100%)**	**68,954 (100%)**

*CSO Annual Report on Vital Statistics 2013

*** Termination of Pregnancy for Fetal Anomaly:** Abortion is illegal in Ireland. However, women in Ireland have the option to travel outside of Ireland for termination of pregnancy following prenatal diagnosis. Where information is available to the registry about these cases, they are included.

Table 5 Infant Birth weight Cork & Kerry Register 2013

Birthweight	No (%) infants in Cork & Kerry Register in Single-ton	No (%) of infants nationally*
Under 2499g	46 (14%)	3835 (6%)
2500g & over	261 (83%)	64912 (94%)
Not stated	9 (3%)	207 (0%)
Total	**316 (100%)**	**68,954 (100%)**

*CSO Annual Report on Vital Statistics 2013

See tables 6 and 7 for prevalence and aetiology of congenital anomalies registered in 2013 in our region.

Table 7 Cases and prevalence per 10,000 births from Cork & Kerry Registry data compared to EUROCAT Full Member Registry data, 2013

(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 (95% CI)	EUROCAT Cases	EUROCAT Prevalence (95% CI)
All Cases	**336***	**350.40 (313.96 - 389.92)**	**8417**	**266.44 (260.78 - 272.19)**
Anomaly				
Nervous System	24	25.03 (16.03 - 37.24)	943	29.85 (27.98 - 31.82)
Eye	5	5.21 (1.66 - 12.21)	170	5.38 (4.60 - 6.25)
Ear, face and neck	0	0.00	57	1.80 (1.37 - 2.34)
Congenital heart defects	93	96.99 (78.29 - 118.80)	2644	83.69 (80.54 - 86.95)
Respiratory	2	2.09 (0.20 - 7.61)	128	4.05 (3.38 - 4.82)
Oro-facial clefts	20	20.86 (12.73 - 32.22)	442	13.99 (12.72 - 15.36)
Digestive system	7	7.30 (2.91 - 15.07)	620	19.63 (18.11 - 21.23)
Abdominal wall defects	5	5.21 (1.66 - 12.21)	167	5.29 (4.52 - 6.15)
Urinary	24	25.03 (16.03 - 37.24)	1116	35.33 (33.28 - 37.46)
Genital	15	15.64 (8.74 - 25.81)	641	20.29 (18.75 - 21.92)
Limb	175	182.50 (156.48 - 211.62)	1532	48.49 (46.10 - 50.98)
Chromosomal	46	47.97 (35.13 - 63.98)	1221	38.65 (36.51 - 40.88)

*NOTE: Table extracted from the EUROCAT website data prevalence reports on Nov 24th 2016. The total number of cases are an underestimate of the true Cork & Kerry Registry cases for 2013 (n=340) because this prevalence data table is based on data submitted to Eurocat on 20/10/2016 and this table has yet to be updated.

EUROCAT 2013 Data is not available for the following years/registries: Austria Styria, Belgium Hainaut, Croatia Zagreb, France Auvergne, Germany Mainz, Ireland Dublin, Norway, Poland Wielkopolska.

Table 7 Aetiology of Congenital Anomaly 2013 Cork & Kerry

Aetiology	No. Cases	%
Isolated cardiac	45	13.20%
Genetic syndrome, skeletal dysplasia and monogenic disorder	17	5.00%
Chromosomal	47	13.80%
Isolated other	185	54.40%
Multiple anomalies	22	6.50%
NTD isolated	10	2.90%
Isolated renal	11	3.20%
Teratogenic syndromes (CMV)	2	0.60%
Poorly specified case	1	0.30%
Total	**340**	**100%**

A word of thanks to the staff of Cork & Kerry Hospitals who facilitate our Research Nurse in 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