Feidhmeannacht na Seirbhíse Sláinte
Health Service Executive

Congenital Anomalies Cork & Kerry

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Cork and Kerry Congenital Anomaly Registry: 2001 data

There were 7933 births (4120 males; 3813 females) in Cork and Kerry in 2001. The number of babies with a birth defect was 227 (2.9%), 216 singleton and 11 twin deliveries. There were 117 male and 109 female infants born with a birth defect. One infant had indeterminate sex.

The age of the mothers who gave birth to a child with a congenital anomaly in 2001 is shown in Table 1. It shows that the prevalence of congenital anomaly increases with increasing maternal age after 35 years of age.

Table 1:

Age Group	All Births	Births with congenital anomalies	Rate/1000 Livebirths & Stillbirths
15-19	325	7	21.54
20-24	1099	34	30.94
25-29	1847	47	25.45
30-34	2757	66	23.94
35-39	1596	65	40.73
40-44	265	8	30.19
45+	13	0	0.00
N/A	31	0	0.00
Total	7933	227	28.61

In 5 (14%) of the stillbirths registered in Cork and Kerry in 2001, a congenital anomaly was present. Stillbirths accounted for 0.5% of total births.

The diagnosis of a congenital anomaly in a child can be delayed. Most (76%) are diagnosed at or shortly after birth. Another 12% will be diagnosed by the time a child is a year old. But for another 12% the diagnosis is made after the age of one year (Table 2).

Table 2:

When Discovered	No.	%
Prenatal diagnosis	24	10.6
At birth	73	32.2
Less than 1 week	75	33
1-4 weeks	5	2.2
1-12 months	22	9.7
Over 12 months	3	1.3
At post mortem	1	0.4
Not entered	8	3.5
Unknown	16	7.1
Total	227	100

A baby with a congenital anomaly is more likely to be born prematurely and to have a low birthweight (Table 3 & 4).

Table 3: Gestational age of infants

Length of gestation	No. of infants in Cork & Kerry Registry (%)	*No. of infants nationally (%)
Under 37	39 (17%)	3101 (5%)
37 & over	188 (83%)	54258 (94%)
Not stated	0	495 (1%)
Total	227	57854

Chi square test: p is ≤0.001. The distribution is significant.
**Source: Report on Vital Statistics, CSO, 2001*

Table 4: Birthweight of infants

Birthweight	No. of infants in Cork & Kerry Registry (%) in singleton	*No. of infants nationally (%)
Under 2500g	27 (12.5%)	2911 (5%)
2500g & over	189 (87.5%)	54453 (94%)
Not stated	0	490 (1%)
Total	216	57854

Chi square test: p is ≤0.001. The distribution is significant.
**Source: Report on Vital Statistics, CSO, 2001*

Table 5: Cases and prevalence per 10,000 births from Cork and Kerry Registry data compared to Eurocat Full Member Registry data, 2001 (includes live births, foetal deaths, and induced abortions where data is available)

Congenital Anomaly	Cork & Kerry Cases	Cork & Kerry prevalence	Eurocat cases	Eurocat prevalence
All cases	227	284.85	14215	224
Anomaly				
Nervous System	22	27.61	1367	21.54
Eye	7	8.78	301	4.74
Ear	5	6.27	232	3.66
Congenital Heart Disease	80	100.39	4133	65.13
Cleft lip with our without palate	13	16.31	564	9.12
Cleft palate	4	5.02	370	5.98
Digestive System	10	12.55	889	14.01
Internal urogenital system	10	12.55	1921	30.27
External Genital system	9	11.29	932	14.69
Limb	47	58.98	2171	34.21
Musculoskeletal and connective tissue	25	31.37	1470	23.16
Chromosomal	23	28.86	2264	35.68
Anomalies outside the normal range*	18	22.59	531	8.37
Total	**273**		**17145**	

**Anomalies outside the Q chapter in ICD10*

Prenatal Screening Policies in Europe

A recent EUROCAT report documents the differing prenatal screening policies in the EU. It notes that the factors that influence prenatal screening policy in a country include resources available, the country's termination of pregnancy law and social and cultural factors. Overall, since the set up of EUROCAT there has been an increase in the proportion of anomalies diagnosed prenatally.

Eleven EU countries reported on their prenatal screening policy under the headings of screening for Down syndrome; indications for prenatal cytogenetic diagnosis; screening for structural anomalies by ultrasound screening; and on termination of pregnancy for foetal anomaly.

Most countries, Ireland and Portugal excepted, provided screening for Down syndrome free of charge to those at increased risk. Screening was available at a cost on request for other mothers. The definition of those at increased risk varied between countries but included an age criterion of maternal age over 35/38 years. Screening was not mandatory.

The indication for prenatal cytogenetic diagnosis was similar to screening for Down syndrome. In the Netherlands, Denmark, Norway and Ireland those defined as being at risk of having a baby with a structural anomaly might be offered an anomaly scan at 18-20 weeks but there is incomplete coverage and regional variation. All other countries surveyed provide at least one anomaly scan for all women at 20 weeks gestation.

Termination of pregnancy was allowed in all the countries surveyed other than in Ireland. France and Portugal allowed termination up to term for lethal conditions. In other countries there was a gestational age upper limit for termination of pregnancy. For foetal anomaly, this upper limit varied from 10 to 24 weeks gestation. It was permissible later in pregnancy subject to the approval of a multidisciplinary committee in a number of countries.

The focus of the report is narrow. The issue of congenital anomaly prevention and control is reviewed only in the context of termination of pregnancy. Prenatal diagnosis of some anomalies such as transposition of the great arteries can allow for the organisation of prompt treatment which may improve outcome. However despite its shortcomings the report is welcome in highlighting this important area of practice.

[1]Boyd P, de Vigan C, Garne E, editors. Prenatal Screening Policies in Europe. N Ireland: EUROCAT, University of Ulster; 2005 http://www.eurocat.ulster.ac.uk/pdf/Special-Report-Prenatal-Diagnosis.pdf

Gastroschisis

Gastroschisis, an abdominal wall defect more prevalent in young mothers, is known to be increasing in prevalence despite the general decrease in the proportion of births to young European women. There have been two clusters of babies born with gastroschisis in the Welsh valleys. The mothers were noted to have poor nutrition with a low maternal body mass index (BMI). No other common factor was found.

A EUROCAT study shows geographical differences within Europe, with the United Kingdom having higher rates of gastroschisis especially among young mothers <20 years, and Italy having a low prevalence. An International Clearing House for Birth Defects (ICBDMS) study found that prevalence rates for gastroschisis were high and increasing in the US, Canada, Scandinavian countries, UK, South America, Mexico and France. Low rates were found in the Mediterranean countries and in Belgium, Austria, and Germany. The study found that prevalence rates for European countries correlated with latitude.

A literature review on the outcome for babies born with gastroschisis found that 90% reported normal health or better. At 5 years of age average growth was found. Fifteen percent of the children continued to have bowel problems but many were mild. Twenty four percent of the babies went on to have further surgery after the neonatal period. The surgery was mostly for adhesions. Some babies experienced other bowel problems, failure of rotation or atresia. Bowel problems were found to decrease over time. Tragically some children survived the neonatal period only to succumb later. On the whole there is a favourable prognosis. Outcome depends on the condition of the bowel at birth.

Congenital Anomaly Registry (CAR) Review

A strategic review of the Cork and Kerry register was done this year. CDC guidelines were used as a framework for the review. A survey of local stakeholders accompanied the first report on data from the Cork and Kerry Congenital Anomaly Register in 2004. The internal strategic review involved a literature review, review of local, national and EUROCAT documents on operation of the register, and semi-structured interviews with key informants. Strong support for the register was expressed in the survey with 98% agreeing it was useful in Cork and Kerry. More agreed that it was useful in the areas of control and prevention and in research than in service planning or in response to public concerns. The greatest local public health use of the data is in the investigation of reports of alleged clusters. The national extension of congenital anomaly surveillance is a public health goal. It is needed to evaluate services and health promotion activities such as folic acid food fortification. Those surveyed favour a national body that builds on and coordinates current registries to maintain the local focus. Strategies to deal with the backlog in data continue. Recommendations to provide regular newsletters and to set up a local steering group have been acted on.