MALTA CONGENITAL ANOMALIES REGISTRY

CONGENITAL ANOMALIES REPORT- 1993 TO 1997

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INTRODUCTION

MALTA CONGENITAL ANOMALIES REGISTER

This is the first in a series of regular reports on congenital anomalies to be issued by the Malta Congenital Anomalies Registry. The register of congenital anomalies was first started in 1983 with data being collected from St. Luke's Hospital as part of a research project funded by the University of Malta. In 1997, the running of the register was assumed by the Department of Health Information and a computerised database was developed with computerised data being backdated to 1993. The register presently collects data from all hospitals on the Maltese Islands and utilises multiple sources of information including information from maternity wards at SLH, Doctors notifications, National Obstetric Information Systems database, Cardiac lab. records, Hospital Activity Analysis database, information from the genetics clinic and others.

The aims of the Malta register are:

- to collect data about all fetal deaths and infants with a diagnosis of congenital anomalies on the islands of Malta and Gozo;
- to detect any changes in occurrence of congenital anomalies;
- to keep a register of all cases of congenital anomalies diagnosed until one year of age;
- to provide data which may be required for epidemiological studies;
- to issue regular reports and provide physicians and the general public with information they may need, always respecting strict confidentiality.

The Malta Congenital Anomalies Registry issues regular six-monthly reports which are widely distributed to all medical practitioners, interested departments and organisations. The aim of these reports is not only to provide statistical information but also to discuss particular topics of interest in the field of congenital anomalies.

The registry also compiles ad hoc Department of Health Information (DHI) Fact Sheets, participates in conferences and responds to specific requests related to congenital anomalies made by parliament, interested individuals, organisations or students.

The Malta Congenital Anomalies Register also has a web site address at:
http://www.magnet.mt/services/health/mcar1.
.

International relationships:

European Registration of Congenital Anomalies (EUROCAT)
The Malta Congenital Anomalies Register is a full member of the EUROCAT programme (http://www.lshtm.ac.uk/php/eeu/eurocat/eurocat.htm), regularly transmitting anonymised data to this programme.

The EUROCAT project is a programme supported by the European Union, represented by the Commission of the European Community for the epidemiologic surveillance of congenital anomalies in Europe. The main objectives are to detect and investigate trends in the frequency of congenital anomalies that could be due to environmental teratogens or mutagens and to evaluate the effectiveness and efficiency of neonatal and perinatal health services.

As congenital anomalies have a relatively low prevalence and good quality exhaustive data is expensive and difficult to collect, a standard European system allows countries using data from regional registries to pool their data for studies and to exploit their differences by comparing them. All registries considered as full members of EUROCAT follow standardised guidelines as defined by the programme. EUROCAT follows the general principles set out by the Assembly of the European Science Foundation (1980 and 1985) concerning the protection of privacy and use of personal data for research[1].

The EUROCAT programme started as a concerted action in 1979. Until 1991, EUROCAT was funded by the EC-DG XII (Directorate General for Science, Research and Development Joint Research Centre, Medical Research Division) and from 1991 to 1998 by the EC-DG V (Directorate General for the Employment, Industrial Relations and Social Affairs, Health and Safety Directorate). Since then local registries and the Scientific Institute of Public Health have been participating in the management of EUROCAT. Surveillance is based on a network of regional registries co-ordinated by a Central Registry which, as from January 2000, is situated in the Environmental Epidemiology Unit, Department of Public Health and Policy, London School of Hygiene and Tropical Medicine in London, UK. Presently there are over 40 regional registries in Europe participating in EUROCAT, either as full or associate members.

The specific objectives of the EUROCAT registration are:

- to provide baseline epidemiologic information on congenital anomalies in Europe;
- to detect and investigate trends in the frequency of congenital anomalies in order to assess the impact of known or suspected risk factors particularly related to exogenic agents, as drugs and the environment, including occupational exposure;
- to evaluate the effectiveness and efficiency of health services (primary prevention, PND and termination of pregnancy, treatment);
- to provide a well documented data base for aetiologic and clinical research;
- to act as information centre which could respond to specific needs, such as the assessment of the impact of environmental accidents or change, or the suspicion of teratogenic influences from food, drugs, or other exposures.

International Clearinghouse of Birth Defects Monitoring Systems (ICBDMS)

The Malta Congenital Anomalies Register is presently applying for associate membership to the ICBDMS; this application is to be approved in September 2000.

The ICBDMS (http://www.icbd.org/index.html) is a non-governmental organization in official relations with the World Health Organization representing more than 30 malformation monitoring programs worldwide. Member programs are actively engaged in the systematic collection and analysis of data for the comprehensive monitoring of congenital malformations.

The organization was established in 1974, at a meeting in Helsinki, Finland, where representatives of malformation monitoring systems in ten countries were present. The mission of the Clearinghouse is to help identify and to prevent birth defects and the spread of an epidemic of congenital malformations by serving as an early warning system. To accomplish this the Clearinghouse has three main objectives:

- exchange of routine information in the prevalence of congenital malformations;
- collaborative epidemiologic research;
- expert consultation and assistance for existing monitoring systems to investigate outbreaks and to establish new monitoring systems.

World Health Organisation (WHO)

Aggregated data from the Malta Congenital Anomalies Register is regularly sent to WHO regarding specific anomalies for inclusion in their datasets.

Projects

The Malta Congenital Anomalies Registry is currently involved in two international studies being co-ordinated by the ICBDMS. These are:

Prevention strategies based on periconceptional folic acid supplementation
This project is being funded by a grant from the CDC (Centres for Disease Control) and is being co-ordinated by the International Clearinghouse for Birth Defects.
The objectives of this International study are to systematically collect and analyze data from countries world-wide in order to:
1) describe and compare public health policies regarding the use of folic acid or multivitamin supplements for the primary prevention of neural tube defects and other birth defects, and to investigate the reasons why different countries, working from the same published data, adopted different policies (or none);
2) evaluate to what extent current public health policies are being implemented and explore the factors involved in the compliance and acceptance of such policies;
3) compare the prevalence of selected birth defects possibly preventable by folic acid supplementation such as neural tube defects, oral clefts, cardiac defects, and limb deficiencies, before and after initiation of folate prevention strategies, and to determine whether different strategies are associated with variations in the prevalence of these defects.

International study of sex ratio of malformed infants
Congenital malformations have still not been extensively studied with regard to the sex distribution. The purpose of this study is to measure the sex ratio of major malformations by registry and by year and to compute the sex ratio for these malformations.

Because the shifts in sex ratio, at least in the general population, are relatively slight, large numbers are needed in order to reach statistically valid results. The Clearinghouse offers the opportunity of a large sample size, allowing the study of sex ratio for specific malformations.

Progress regarding the above two studies will be reported in the six-monthly reports issued by the registry.

PART ONE -

DEFINITIONS AND METHODS

THE STUDY POPULATION

The Malta Congenital Anomalies Register is population based and covers all births on the islands of Malta and Gozo which amount to just under 5000 births per year. All infants, until one year of age, who are diagnosed or suspected of having a congenital anomaly until one year of age are included.

The small size and population of the islands (Area: 316 km^2, Population: 376,513 in 1997); the geographically well defined boundaries, absence of significant ethnic minority groups and illegality of termination of pregnancy make the islands ideal for epidemiological studies.

This report covers all births diagnosed with congenital anomaly from 1993 to 1997 and registered at the Registry by December, 1999. Denominator data of total livebirths and stillbirths were obtained from the relevant Demographic Review of the Maltese Islands[2].

ASCERTAINMENT METHODS

To ensure as complete an ascertainment as possible, the Malta Congenital Anomalies Register makes use of active case finding and multiple sources of information. Cases of congenital anomalies diagnosed in the first few days of life in babies born at St. Luke's Hospital are recorded by staff from the registry who visit the postnatal wards daily. Gozo General Hospital and private hospitals (St. Philip's Hospital, Capua Palace Hospital and St. James' Hospital) notify any cases of congenital anomalies using a standardised report form (Annex 1). All paediatricians, paediatric surgeons and other medical doctors are encouraged to report any infants with congenital anomalies under their care to the registry. Other sources of information used in this report include: Echocardiography Lab., Genetics Clinic, St. Luke's Hospital Maternity Systems Database, St. Luke's Hospital Activity Analysis Register, National Mortality Register, Pathology Autopsy reports, Congenital Heart Defects Register, National Cancer Register and the Hypothyroid screening programme. The process of data collection and management is outlined in Figure 1.

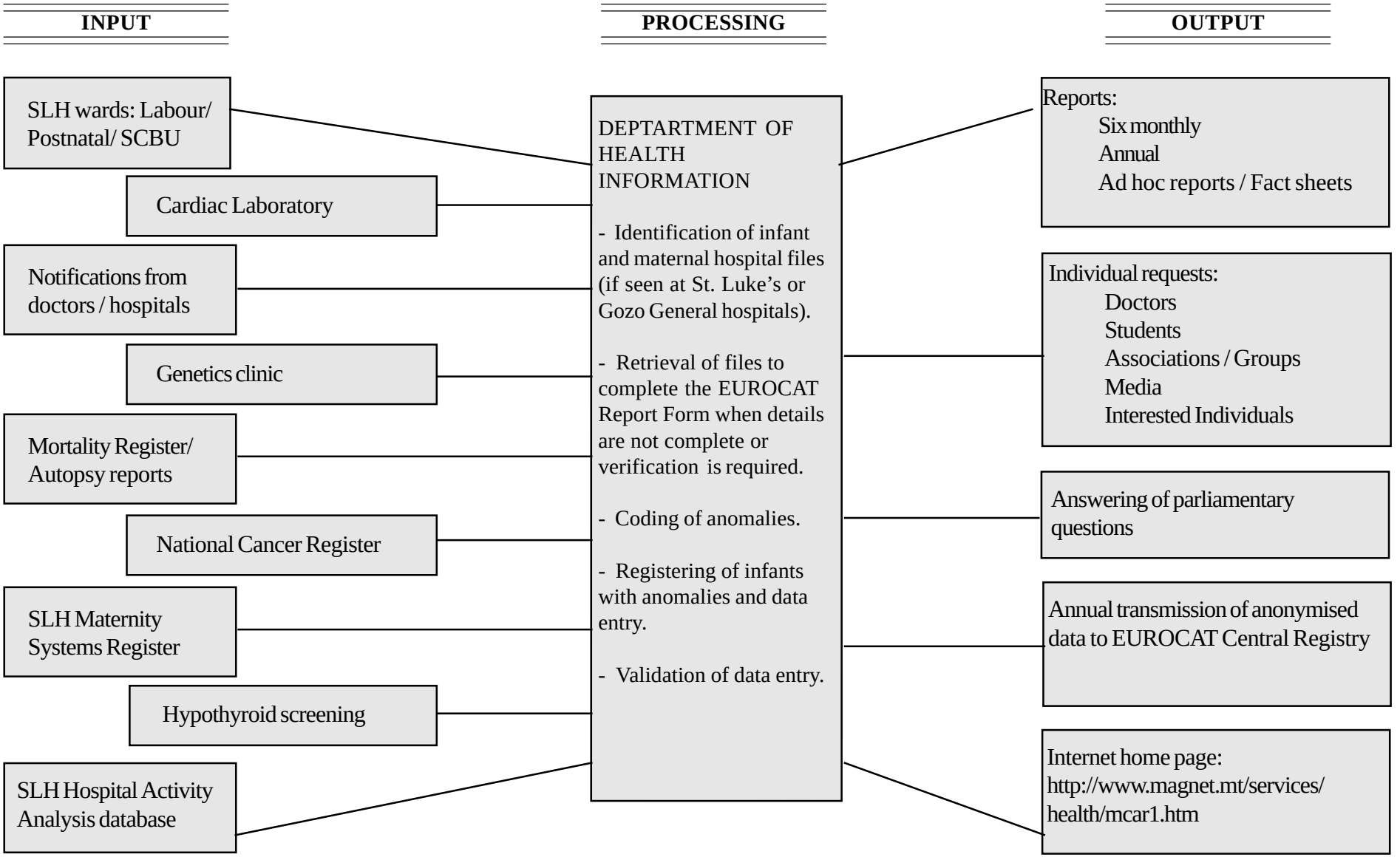
Every effort is made to record all cases of congenital anomalies. However it is extremely difficult, if not impossible, for any registration system to claim complete case ascertainment or complete accuracy of information. First of all, neither ascertainment nor accuracy are absolute in a world of changing diagnostic methods and diagnostic definitions. Furthermore, depending on the malformation, some cases may escape the system.

Ascertainment of livebirths and fetal deaths

Livebirths with congenital anomalies are not always diagnosed at birth or in the early neonatal period, particularly certain cardiac anomalies, internal urogenital system anomalies, central nervous system anomalies, eye anomalies (cataract) and pyloric stenosis for example. For this reason all anomalies diagnosed until one year of age are included in the register, this is inkeeping with EUROCAT guidelines.

Fetal deaths include all registered malformed cases of spontaneous fetal deaths or stillbirths of 20 weeks gestational age or more.

Figure 1 - Data collection and management by the Malta Congenital Anomalies Registry (1993-97)



DEFINITIONS AND EXCLUSIONS

In this report the term 'congenital anomalies' refers to structural defects (congenital malformations, deformations, disruptions and dysplasias), chromosomal abnormalities, inborn errors of metabolism and hereditary diseases. This is the definition given by EUROCAT guidelines for the registration of congenital anomalies.

Minor congenital anomalies are not normally included in congenital anomalies registers as they would markedly swell the incidence figures and dilute the significance of major anomalies. The precise line of demarcation between anomalies to be included and those to be excluded is not clear cut and may be influenced by subjective differences in interpretation. Minor anomalies do not in themselves have serious medical or cosmetic consequences for the child. Some may nevertheless be of certain significance, since they can be predictive of major underlying pathologies. There is increasing awareness of dysmorphic syndromes which present great diagnostic difficulties, not only in their precise identification, but also in their recognition which may be delayed for many months before they are detected.

EUROCAT applies a standard list of minor and commonly occurring anomalies for exclusion; this list is given in Annex 2. These conditions are not registered unless occurring in combination with other major anomalies.

It is necessary to exercise great care and attention to detail in distinguishing some of the anomalies listed for exclusion from those which are to be registered. Thus glandular and coronal hypospadias are excluded wheras penile and perineo-scrotal hypospadias are to be registered. Similarly, structural talipes has to be distinguished from postural talipes which is excluded. Birthmarks are registered as skin anomalies only when they exceed an area of $4cm^2$.

Minor anomalies such as single palmar crease, low set or dysmorphic ears, slanting palpebral fissures, high arched palate, micrognathia, and other dysmorphic features of the face and lips are not included in the register unless associated with other major defects. These anomalies are often difficult to define accurately and interpretation may be highly subjective especially for those who are not experienced in dysmorphology; nevertheless they are highly significant in syndrome identification. Also excluded from the register are hernias, varicocele, undescended testes, spina bifida occulta and pre-auricular pits or tags.

CASE INFORMATION

For each baby with an anomaly, information about the child, the diagnosis, the pregnancy, the parents, their occupation and risk factors (assisted conception, illness before pregnancy, habitual and unusual exposures, drugs), diagnosis of malformations, and family history is recorded. Annex 4 shows the details recorded on the EUROCAT Report Form. Case records are updated when necessary (e.g. diagnosis of later discovered anomalies, change or precision in diagnosis, knowledge of an additional risk factor).

CODING OF CONGENITAL ANOMALIES

The Malta Congenital Anomalies Register codes anomalies using a six-digit EUROCAT code, which is a compatible expansion of the coding system of the British Paediatric Association Classification of diseases (BPA-9)[1]. The BPA-9 is itself a five-digit extension of the 9th revision of the International Classification of Diseases. Since 1998, anomalies have been coded using the Royal College of Paediatrics and Child Health (RCPCH) Classification of Diseases which is a paediatric adaptation of ICD-10 (International Classification of Diseases - 10^{th} Edition).

Each anomaly is precisely and individually coded by a medical officer at the registry. As far as possible all case histories are reviewed to ensure that all details are included in the registration form.

For reporting purposes, EUROCAT classification subgroups are defined[4]. These subgroups make the interpretation of results more meaningful for the clinician and facilitates comparisons of prevalence rates between centres in the EUROCAT network. A list of EUROCAT classification subgroups is given in Annex 3**.**

CALCULATION OF PREVALENCE RATES AND STATISTICAL METHODS

Since some babies are born with more than one anomaly, this report makes a clear distinction between the analyses of the numbers of infants with one or more congenital anomalies and the analyses of the numbers of individual congenital anomalies. As some infants may have more than one anomaly, the number of anomalies do not add up to the number of infants.

When counting infants, each baby is considered once irrespective of how many anomalies he/she may have. Therefore, the *prevalence of infants/fetuses with congenital anomalies* is expressed as the number of infants/fetuses registered with one or more congenital anomalies divided by the total number of births - live, still or induced abortions in that period.

$$\text{Prevalence of infants/fetal deaths with congenital anomalies*} = \frac{\text{Number of infants/fetal deaths registered with anomalies}}{\text{Total number of infants/fetal deaths in that period}}$$

When counting anomalies, babies with more than one anomaly within a specific subgroup/ICD classification group are counted only once within that class. Babies with anomalies affecting more than one specific subgroup/classification group are counted once within each class. This method of counting anomalies is that adopted by EUROCAT Central Registry for reporting purposes.

The *total prevalence rate of congenital anomalies* is expressed as the number of anomalies registered in livebirths/fetal deaths divided by the total number of livebirths and fetal deaths in that period.

$$\text{Total prevalence rate of congenital anomalies*} = \frac{\text{Total number of anomalies registered in livebirths/fetal deaths with anomaly}}{\text{Total number of livebirths/fetal deaths in that period}}$$

The *livebirth prevalence rate of congenital anomalies* is expressed as the number of liveborn affected babies divided by the number of livebirths in that period.

$$\text{Livebirth prevalence rate *} = \frac{\text{Total number of anomalies registered in livebirths}}{\text{Total livebirths in that period}}$$

* Expressed per 10,000 births

Whenever prevalence rates of other countries were considered, the number of induced abortions due to prenatal diagnosis of congenital anomaly were included with the number of fetal deaths due to anomaly. This is not applicable to Malta since termination of pregnancy is illegal.

Statistical interpretations were performed using the statistical packages Epi Info 6 and SPSS.

PART TWO -

SUMMARY ANALYSIS

Summary Analysis
Due to the relatively small number of cases of congenital anomalies registered annually, interpretation of data for one particular year may be unreliable. It is therefore important to analyse aggregated data for several years for more meaningful interpretation. This will minimise errors due to the fluctuation of small number statistics.

Prevalence
In 1997 there were 4,864 total births (live and fetal deaths) on the Maltese Islands; of these 203 (4.2%) babies were registered as having one or more congenital anomalies giving a prevalence of 41.8 per 1,000 total births. Between 1993-1997 there were 902 babies (482 males, 418 females and 2 with indeterminate sex) registered out of a total of 24,510 births. The overall prevalence of babies registered as having one or more major congenital anomalies from 1993-1997 was therefore 36.8 /1,000 total births.

Fetal deaths
During the year 1997, 4 out of the 203 babies registered with anomalies (2.0%) were fetal deaths whereas only 29 out of the 4864 total births (0.6%) on the islands were fetal deaths. Using data from 1993-1997 this greater proportion of fetal deaths in babies with anomalies (22 out of 902 cases - 2.4%) when compared to that for all babies (145 out of 24,510 births - 0.6%) is also apparent (Table 4). The difference is statistically significant.

Gender distribution
Persistently more male babies are registered as having one or more congenital anomalies; in 1997 there were 111 (55%) male babies registered while there were 91 (45%) female babies and one baby with indeterminate sex. For 1993-1997 these percentages were 53% and 46% respectively and two babies were of indeterminate sex.

Table 1- Gender distribution of babies registered as having an anomaly 1993-1997

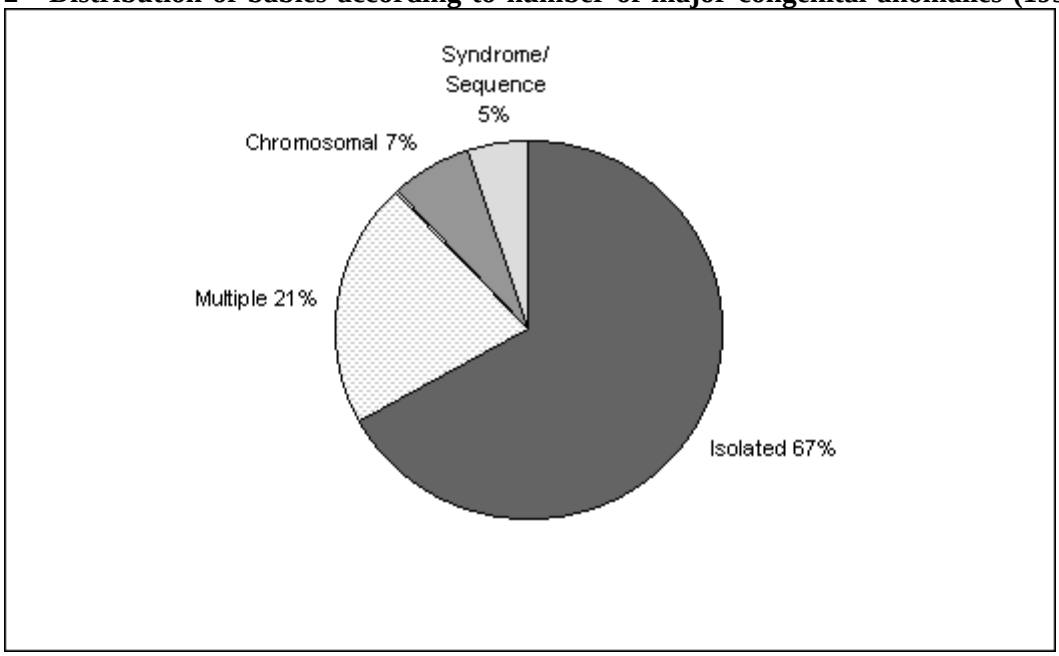
	Total births	Births registered as having an anomaly	Proportion of total births with anomaly /1000
Male	12731	482	37.8
Female	11777	418	35.4
Indeterminate sex	2	2	-
Total	**24510**	**902**	**36.8**

In spite of there being more male babies registered as having an anomaly, the difference in proportions is not statistically significant.

Isolated vs. Multiple anomalies
The majority of babies registered with congenital anomalies have isolated defects. In 1997, out of 203 babies registered with congenital anomalies, 145 (71%) had isolated defects, 16 (8%) had recognised syndromes or sequences and 42 (21%) had multiple unrelated anomalies.

Analysing data from 1993-1997 similar patterns were seen with 607 (67%) of the registered babies being reported to have isolated anomalies and 193 (21%) had multiple unrelated anomalies. 60 (7%) had chromosomal anomalies and 42 (5%) had multiple anomalies recognised as a syndrome or sequence (Figure 2).

Figure 2 - Distribution of babies according to number of major congenital anomalies (1993-97)



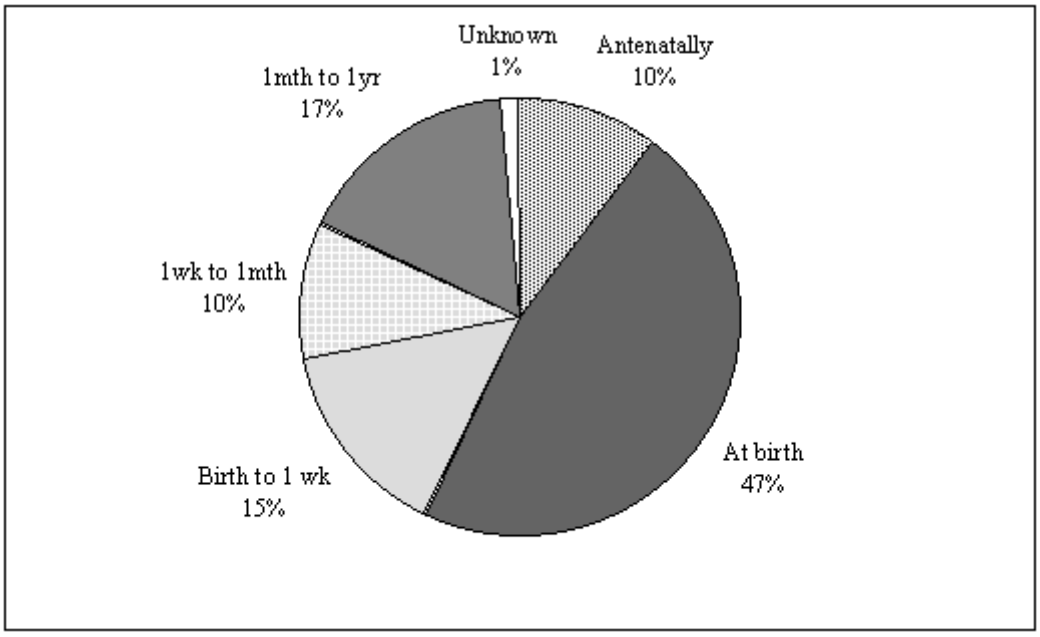
Source: Table 8

Time of diagnosis

Most babies with major congenital anomalies are diagnosed at or soon after birth. In 1997, 87% of babies registered with anomalies were diagnosed in the perinatal period (ie. until one month of age).

The figure below shows the distribution of babies with congenital anomalies according to the time of diagnosis. Between 1993-1997, 82% of babies were diagnosed in the perinatal period. A not insignificant proportion (17%) were however diagnosed after 1 month of age and before 1 year of age. This emphasises the importance of the registry to follow up babies throughout the first year of life for purposes of registration of congenital anomalies.

Figure 3 - Time of diagnosis of babies with major congenital anomalies



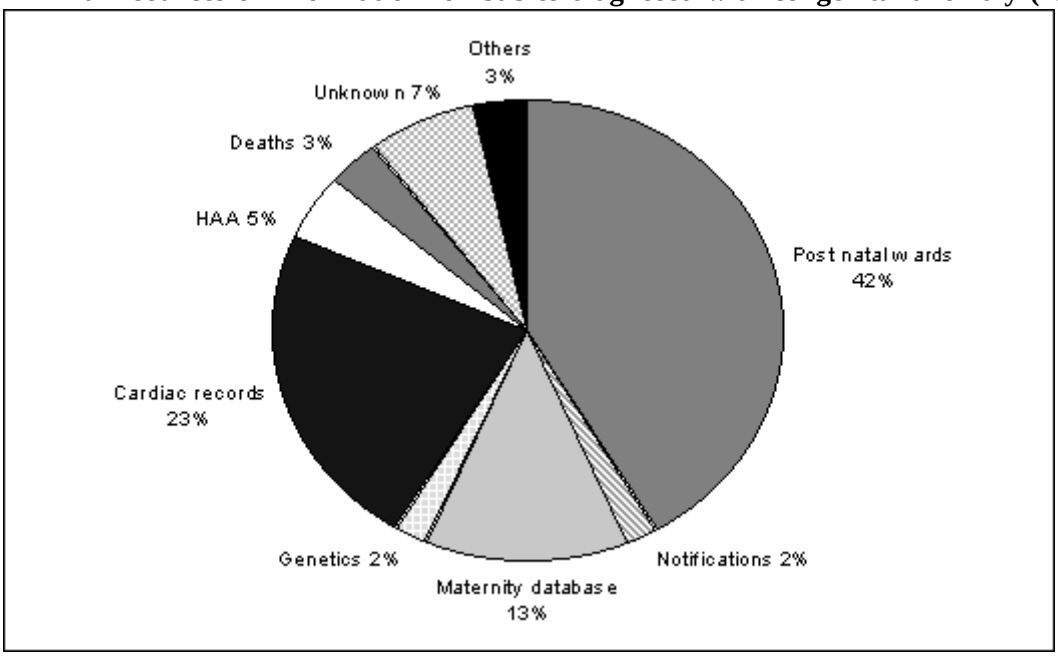
Source: Table 9

Sources of information

Multiple sources of information are employed to identify babies to be registered in the Malta Congenital Anomalies Register. The main sources of information in 1997 were: active data collection from St. Luke's Hospital Maternity Wards (40 %) followed closely by data from Cardiac lab. records and the Congenital Heart Diseases register (38 %).

Between 1993-97, the main source of information was active data collection from maternity wards in St. Luke's Hospital (42%), followed by pick up from Cardiac records including Cardiac Lab and the Congenital Heart Diseases Register (23%). The SLH Maternity Systems database and Hospital Activity Analysis database were the sources of information for 13% and 5% of cases respectively. The National Mortality Register and autopsy records accounted for 3% of cases; both doctor's notifications and genetic clinic records accounted for 2% of cases each and 3% were from miscellaneous other sources. For the remaining 7% of babies registered, the primary source of information was unrecorded.

Figure 4 - Main sources of information for babies diagnosed with congenital anomaly (1993-97)



Source - Table 10

Maternal ages

In 1997, the average maternal age for babies registered with congenital anomalies was 29 years, 35 years for babies with chromosomal defects and 29 years for babies with non-chromosomal defects.

Analysis of 1993-1997 data showed that the average maternal ages of babies with non-chromosomal anomalies were not significantly different from those of all births. However average maternal ages of babies with chromosomal defects were significantly higher than the general population.

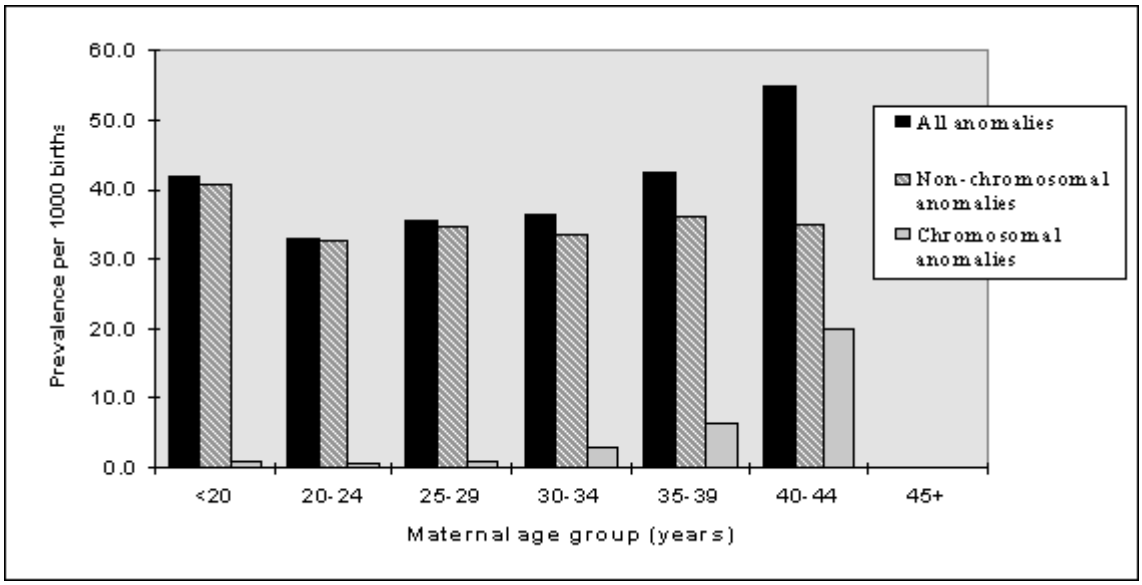
Table 2 - Average maternal ages for all deliveries and deliveries of babies registered with anomalies (1993-97)

	Average maternal age (years)
Deliveries of babies with congenital anomalies (excluding chromosomal)	29
Deliveries of babies with chromosomal anomalies	35*
All deliveries	29

*the difference in this average maternal age is statistically significant from that of all deliveries

Prevalence of congenital anomalies increases significantly in the older maternal age groups. This is due mainly to the increase in chromosomal anomalies occurring in these age groups. In fact when considering non-chromosomal anomalies only, it is seen that there is an increased prevalence in the youngest maternal age group ie. <20years (Figure 5).

Figure 5 - Differences in variation of maternal age groups for all anomalies, non-chromosomal and chromosomal anomalies (1993-97)



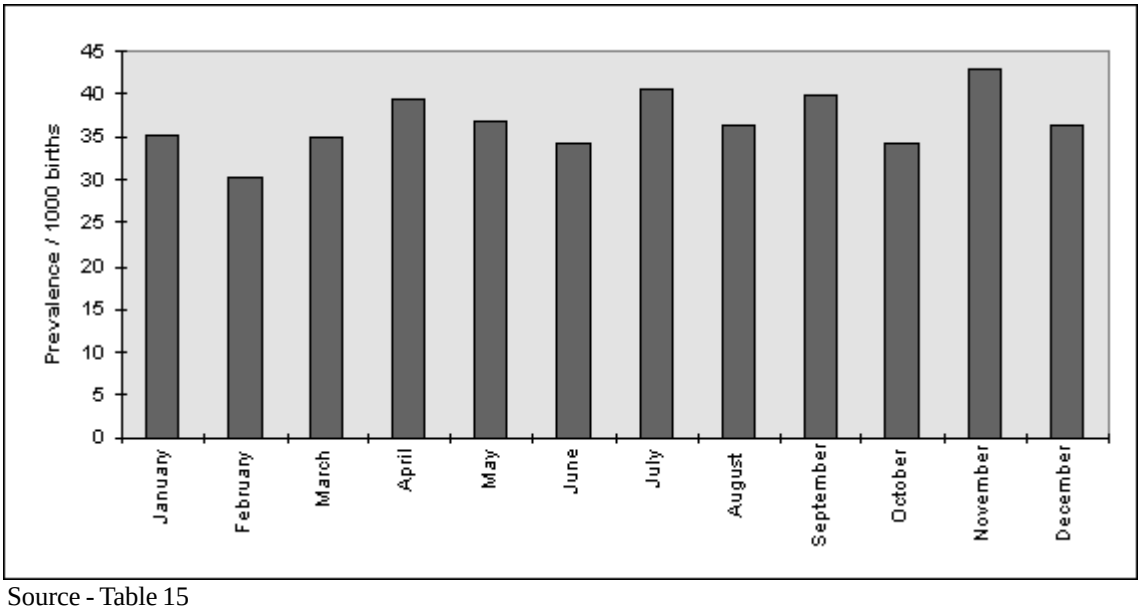
Source: Table 12

Seasonal distribution

The maximum prevalence of babies with anomalies registered in 1997 was in the month of September (61/ 1,000 births) and the least was in the month of December (36.1/1,000 births). The figure below shows the seasonal variation of prevalence of babies registered with anomalies between 1993-1997.

The pattern shows no definite seasonal variation in prevalence of babies with congenital anomalies when considering all anomalies together. The pattern may change if only babies with one particular anomaly only are considered.

Figure 6 - Monthly distribution of babies registered with congenital anomalies (1993-97)



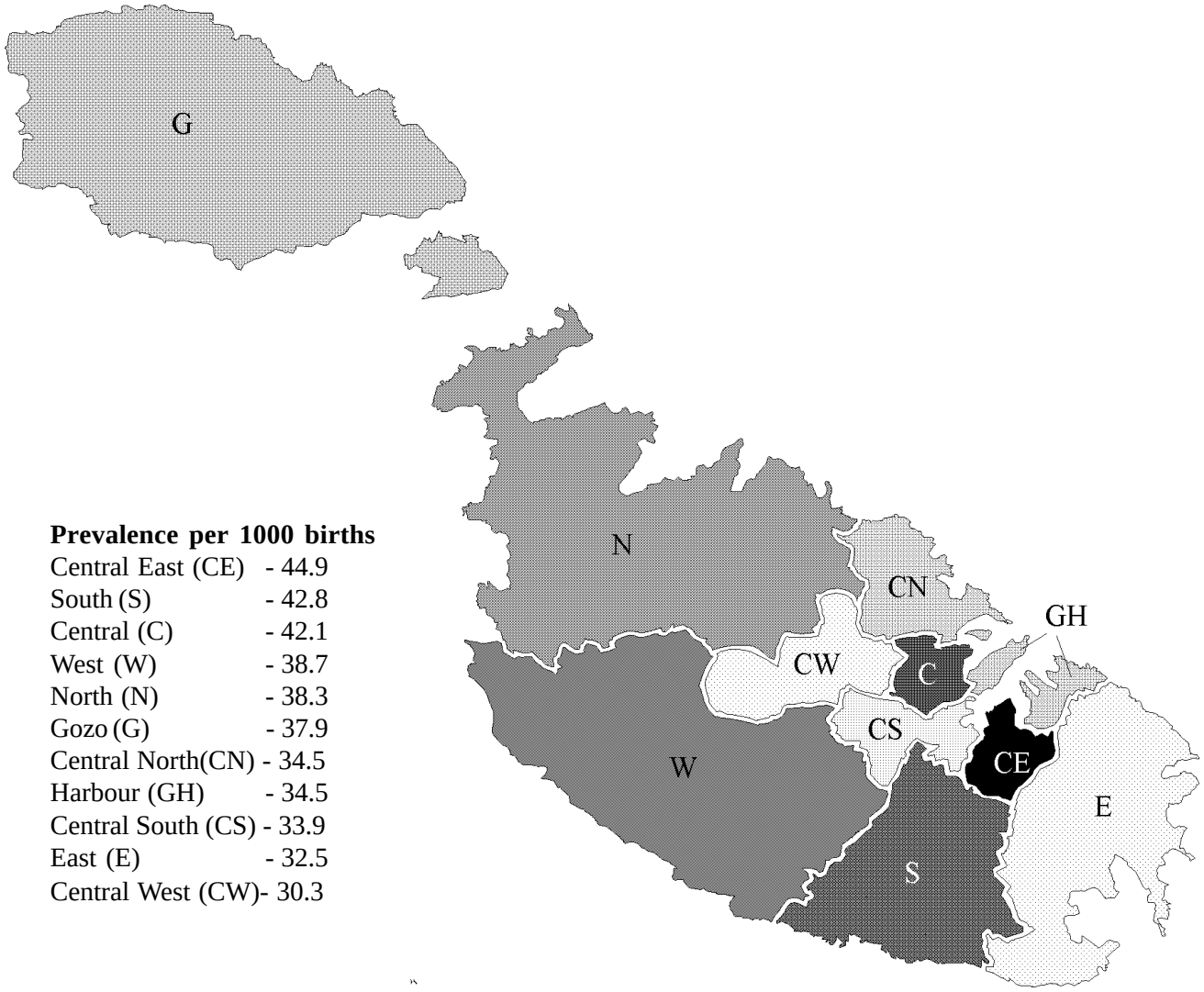
Source - Table 15

Geographic distribution

Malta and Gozo may divided into 11 regions (see Annex 5). In 1997 the highest prevalence of livebirths with congenital anomalies was in the Central East Region (44.9/1,000births) and the lowest was in the Central West (30.3/1,000 births) see Table 16.

The map below shows the distribution of the prevalence of all babies registered with congenital anomalies between 1993-1997. There are no statistically significant differences between the various regions.

Figure 7 - Geographical distribution of all babies registered with congenital anomalies in Malta and Gozo from 1993-1997.



Prevalence per 1000 births

Central East (CE)	- 44.9
South (S)	- 42.8
Central (C)	- 42.1
West (W)	- 38.7
North (N)	- 38.3
Gozo (G)	- 37.9
Central North(CN)	- 34.5
Harbour (GH)	- 34.5
Central South (CS)	- 33.9
East (E)	- 32.5
Central West (CW)	- 30.3

Distribution of anomalies

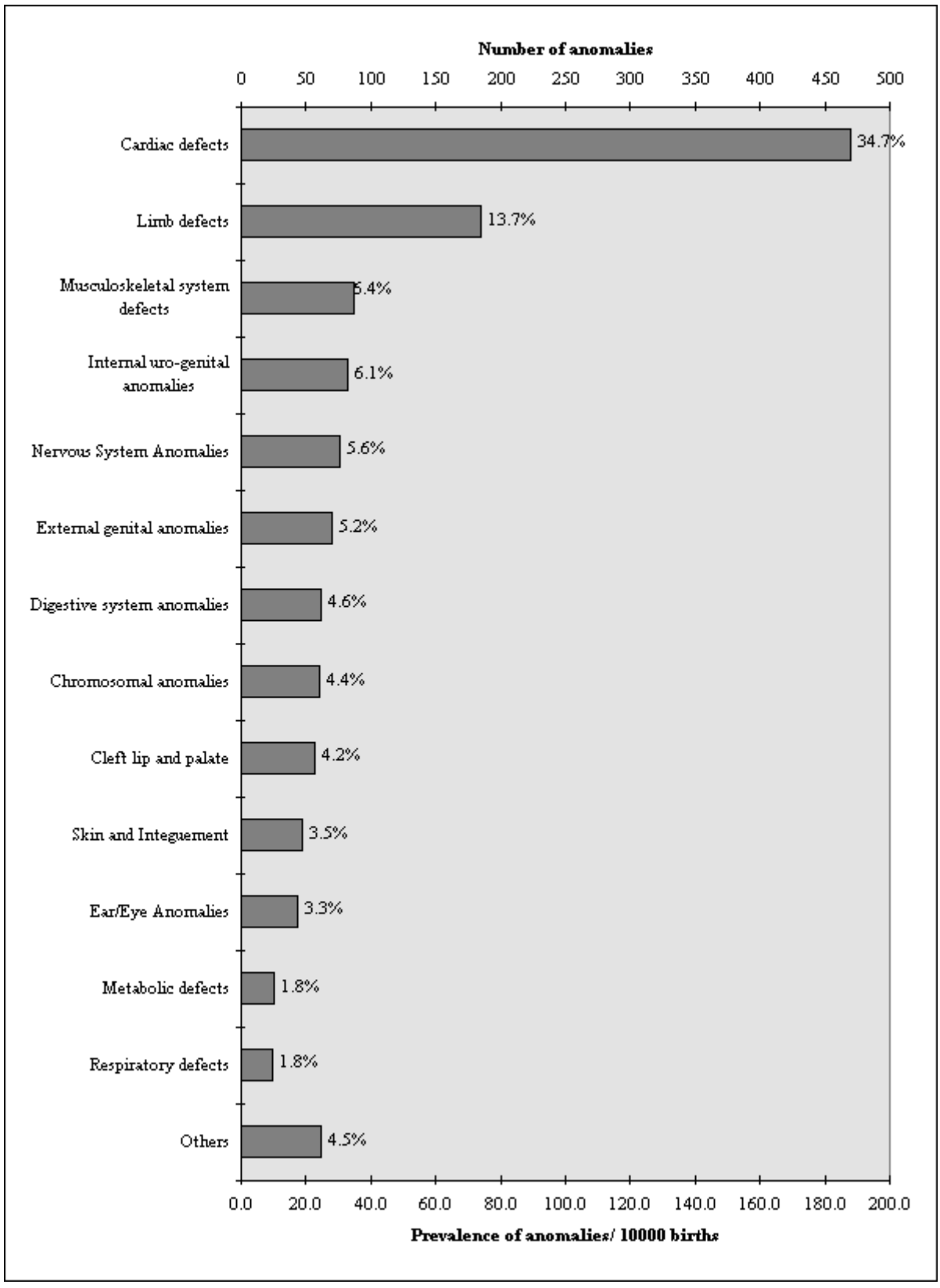
One baby may have one or more different anomalies affecting the same or different systems. The total number of anomalies registered, therefore, does not add up to the same number of babies with anomalies. The table below gives a breakdown of infants/fetuses according to anomaly groups. In this tabulation, a baby having more than one anomaly classified within the same EUROCAT subgroup was counted only once within this group. However a baby/fetus with anomalies in different EUROCAT subgroups was counted once within each subgroup. The distribution of anomalies in decreasing order of frequency is depicted in Figure 8.

The main anomalies encountered in babies/fetuses in 1997 were cardiovascular (37.9%) followed by limb and musculoskeletal defects (12.4% and 7.4% respectively). This pattern is similar to that encountered in previous years.

Table 3 - Distribution of babies/fetuses with major anomalies registered in Malta from 1993-1997 according to the group/system(s) affected

Defect Sub Group *	System(s) affected	1993	1994	1995	1996	1997	1993-97		
							No.	Prev. /1000 births	Rel. Freq. %
01-04	Nervous system anomalies	14	20	16	14	12	76	31.0	5.6
05-06	Ear and Eye anomalies	6	9	11	9	9	44	18.0	3.3
07-09	Cardiac defects	69	102	81	111	107	470	191.8	34.7
10	Respiratory system defects	3	9	4	3	5	24	9.8	1.8
11	Cleft lip and palate	13	12	12	9	11	57	23.3	4.2
12-14	Digestive system anomalies	8	13	12	11	18	62	25.3	4.6
15-16	External genital anomalies	19	11	13	11	16	70	33.5	5.2
17-19	Internal urogenital anomalies	15	23	14	15	15	82	28.6	6.1
20-23	Limb defects	33	40	53	24	35	185	75.5	13.7
25-29	Musculoskeletal defects	17	21	11	17	21	87	35.5	6.4
30-31	Skin and Integuement	13	10	9	6	9	47	19.2	3.5
32-33	Chromosomal anomalies	17	8	14	12	9	60	24.5	4.4
34	Metabolic defects	5	4	7	3	6	25	10.2	1.8
	Others	12	13	12	15	9	61	24.9	4.5
	Total	244	295	269	260	282	1350	552.0	100

Figure 8 - Distribution (%) of infants/fetuses registered in Malta from 1993-1997 according to the group/system affected (Data source: Table 3).



PART THREE -

TABLES

A - ANALYSIS OF INFANTS WITH ANOMALIES

B - ANALYSIS OF ANOMALIES

A - ANALYSIS OF INFANTS WITH ANOMALIES

LIVEBIRTHS AND FETAL DEATHS
(for residents of the Maltese Islands)

Table 4 - Distribution of livebirths and stillbirths in all babies and in babies with anomalies in the Maltese Islands from 1993-1997.

Year	All babies*			Babies registered with anomalies		
	Live births	Fetal deaths	Total	Live births	Fetal deaths	Total
1993	5147	25	5172	159	4	163
1994	4826	37	4863	179	8	187
1995	4613	20	4633	182	1	183
1996	4944	34	4978	161	5	166
1997	4835	29	4864	199	4	203
1993-97	**24365**	**145**	**24510**	**880**	**22**	**902**

Table 5 - Proportion of fetal deaths with a congenital anomaly recorded as underlying cause of death

	1993	1994	1995	1996	1997	1993-97
Fetal deaths*	25	37	20	34	29	145
No. fetal deaths attributed to congenital anomalies*	1	4	0	5	3	13
Percentage (%)	4.0	10.8	0.0	14.7	10.3	8.9

Table 6 - Proportion of babies dying with a congenital anomaly recorded as underlying cause of death from livebirth to 1 week of age

	1993	1994	1995	1996	1997	1993-97
All deaths birth to 1 wk*	24	29	29	38	21	141
No. of these deaths due to congenital anomalies*	12	8	9	8	9	46
Percentage (%)	50.0	27.6	31.0	21.1	42.9	32.6

* Data taken from National Mortality Register

Table 7 - Proportion of babies dying with a congenital anomaly recorded as underlying cause of death after 1 week to 1 year of age

	1993	1994	1995	1996	1997	1993-97
All deaths 1 wk to 1 yr*	19	15	12	15	10	71
No. of these deaths due to congenital anomalies*	7	7	4	7	4	29
Percentage (%)	36.8	46.7	33.3	46.7	40.0	40.8

* Data taken from National Mortality Register

DISTRIBUTION OF INFANTS/FETUSES WITH CONGENITAL ANOMALY ACCORDING TO DIAGNOSTIC CATEGORY

The overall diagnosis of an infant/fetus may be classified as having a

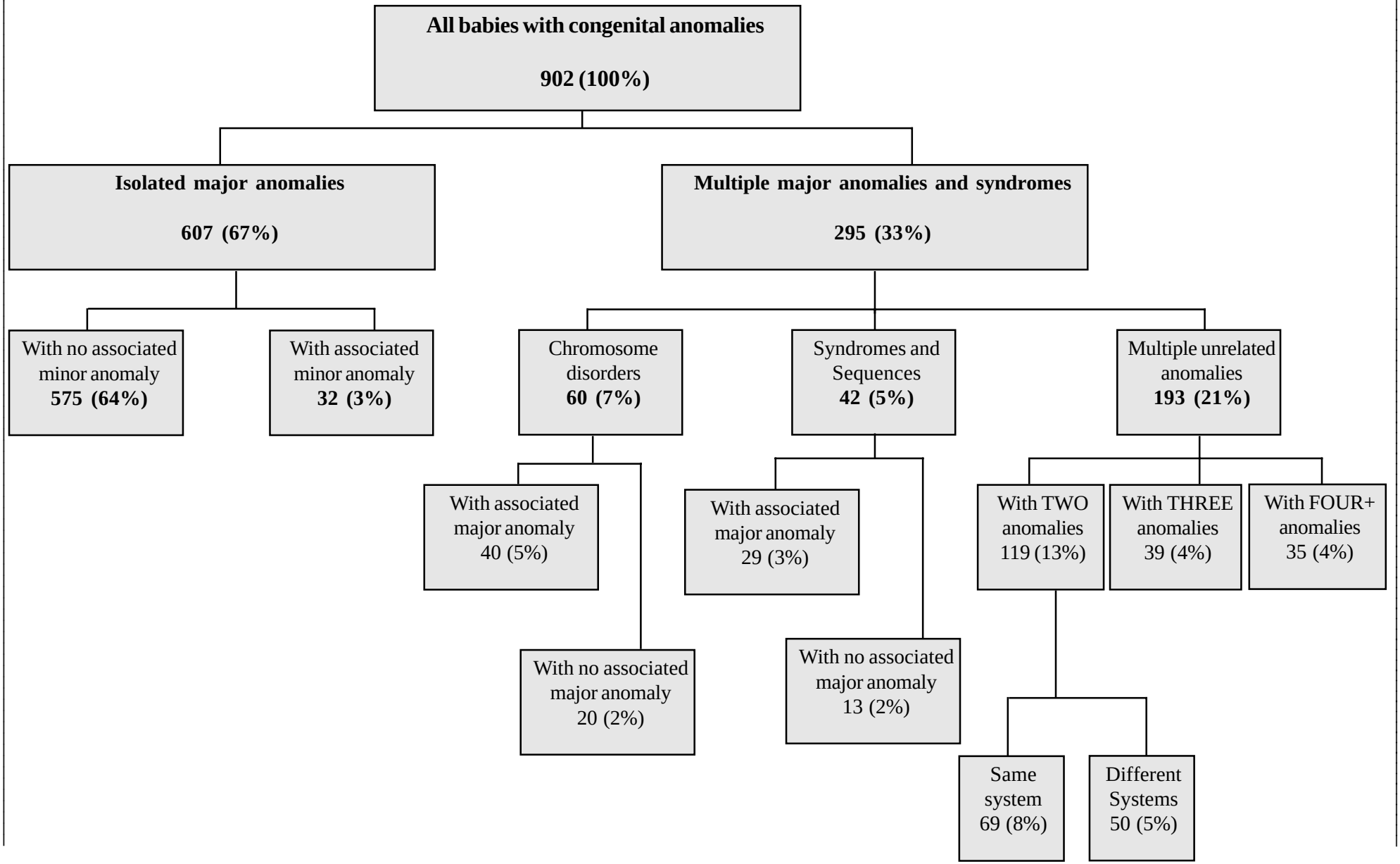
I	Single anomaly	Infant or fetus with an isolated major anomaly
II	Syndromes	Infant or fetus with a pattern of more than one anomaly, recognised as a syndrome, sequence, or metabolic disorder. Recognised associations (with unknown aetiology) are also included here.
III	Multiple Anomalies	Infants or fetuses with more than one anomaly, not recognised as a syndrome or sequence.

Table 8 - Number of registered children according to diagnostic category of the congenital anomalies and per year of birth

ICD-9 Code	Diagnostic category	Year					
		1993	1994	1995	1996	1997	93-97
	I Single anomaly	**107**	**130**	**130**	**95**	**145**	**607**
7400-7420	- neural tube defect	4	6	3	1	2	16
7421-7429	- other central nervous system	1	9	3	2	0	15
7430-7449	- eye, ear, face and neck	2	6	4	9	4	25
7450-7479	- heart and circulatory system	41	46	52	42	63	244
7480-7489	- respiratory system	1	4	2	1	3	11
7490-7493	- oral clefts	9	7	10	4	7	37
7500-7519	- digestive system	5	11	6	4	12	38
7520-7529	- genital organs	15	5	5	5	12	42
7530-7539	- urinary system	3	14	7	3	4	31
7540-7569	- musculoskeletal anomalies	13	16	29	16	22	96
7570-7579	- skin & integuement	5	3	3	0	7	18
	- others	8	3	6	8	9	34
	II Syndromes	**29**	**16**	**21**	**20**	**16**	**102**
7580-7589	- chromosomal	17	8	14	12	9	60
	- other syndromes/ sequences / associations	12	8	7	8	7	42
	III Multiple Anomalies	**27**	**41**	**32**	**51**	**42**	**193**
	– multiple anomalies of one system	17	18	10	32	16	93
	- multiple cardiovascular	*9*	*15*	*7*	*26*	*13*	*70*
	- multiple musculoskeletal	*3*	*3*	*2*	*1*	*1*	*10*
	– multiple anomalies of different systems	10	23	22	19	26	100

This distribution is represented graphically in the following figure.

Figure 9 - Distribution of babies registered with congenital anomalies according to number of anomalies (1993-1997)



DISTRIBUTION OF INFANTS/FETUSES WITH CONGENITAL ANOMALIES BY

TIME OF DIAGNOSIS

Table 9 - Distribution of babies according to time of diagnosis

Time of diagnosis	1993	1994	1995	1996	1997	93-97	%
Pre-natally	13	24	11	19	85	92	10
At birth	86	87	99	65	35	422	47
Within first week of life	15	20	30	35	16	135	15
1 week to 1month	19	27	16	12	41	90	10
1 month to 1 year	29	26	24	30	25	150	17
Uncertain	1	3	3	5	1	13	1.4
Total	**163**	**187**	**183**	**166**	**203**	**902**	**100**

SOURCE OF INFORMATION

Table 10 - Distribution of babies registered with anomaly by first source of information

First source of Information	1993	1994	1995	1996	1997	93-97	%
Active data collection from maternity wards & SCBU	105	99	81	11	81	377	42
Doctors' notification	1	2	2	6	5	16	2
Death Register/Postmortem	3	2	4	16	6	31	3
Cardiac Lab records & Congenital heart disease reg	23	27	33	48	78	209	23
Maternity systems database	0	1	34	70	10	115	13
HAA	1	10	14	6	11	42	5
Genetics clinic	5	3	6	4	2	20	2
SLH operations register	0	0	2	3	6	11	1
Others	4	4	4	2	4	18	2
Unknown	21	39	3	0	0	63	7
Total	**163**	**187**	**183**	**166**	**203**	**902**	**100**

GENDER

Table 11 - Distribution of babies with anomaly (livebirths and fetal deaths) by sex (1993-97)

Sex	Total number of babies with anomalies (Prevalence per 1000 births)											
	1993		1994		1995		1996		1997		93-97	
Males	90	(33.5)	95	(38.0)	108	(44.7)	78	(30.3)	111	(43.3)	482	(37.9)
Females	73	(29.4)	91	(38.5)	75	(33.8)	88	(36.6)	91	(39.5)	418	(35.5)
Indeterminate	-		1		-		-		1		2	
Total	**163**	**(31.5)**	**187**	**(38.4)**	**183**	**(39.5)**	**166**	**(33.3)**	**203**	**(41.7)**	**902**	**(36.8)**

MATERNAL AGE

Table 12 - Distribution of all babies with congenital anomalies according to maternal age

Maternal age (yrs)	Total number of babies with anomalies (Prevalence per 1000 births)					
	1993	1994	1995	1996	1997	93-97
<20	11 (62.1)	5 (32.9)	10 (69.9)	5 (20.9)	9 (36.6)	40 (41.8)
20-24	21 (20.6)	41 (41.8)	33 (37.7)	25 (25.6)	43 (39.9)	163 (33.0)
25-29	53 (28.9)	49 (29.3)	66 (40.3)	64 (36.3)	73 (43.7)	305 (35.5)
30-34	42 (30.2)	54 (39.7)	49 (37.8)	44 (34.4)	48 (41.2)	237 (36.5)
35-39	28 (45.7)	28 (48.9)	20 (35.8)	22 (37.9)	26 (44.4)	124 (42.6)
40-44	8 (62.5)	10 (86.2)	5 (42.7)	6 (47.6)	4 (34.8)	33 (54.8)
45+	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)
Total	**163 (31.5)**	**187 (38.4)**	**183 (39.5)**	**166 (33.3)**	**203 (41.7)**	**902 (36.8)**

n/a - not applicable

Table 13 - Distribution of babies with non-chromosomal defects according to maternal age

Maternal age (yrs)	Total number of babies with anomalies (Prevalence per 1000 births)					
	1993	1994	1995	1996	1997	93-97
<20	11 (62.1)	5 (32.9)	9 (62.9)	5 (20.9)	9 (36.6)	39 (40.8)
20-24	21 (20.6)	40 (40.8)	33 (37.7)	24 (24.5)	43 (39.9)	161 (32.6)
25-29	52 (28.4)	47 (28.1)	66 (40.3)	63 (35.7)	70 (41.9)	298 (34.7)
30-34	37 (26.6)	53 (39.0)	41 (31.7)	41 (32.1)	46 (39.5)	218 (33.6)
35-39	21 (34.3)	25 (43.6)	16 (28.7)	19 (32.8)	24 (41.0)	105 (36.1)
40-44	4 (31.3)	9 (77.6)	4 (34.2)	2 (15.9)	2 (17.4)	21 (34.9)
45+	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)
Total	**146 (28.2)**	**179 (36.8)**	**169 (36.5)**	**154 (30.9)**	**194 (39.9)**	**842 (34.4)**

n/a - not applicable

Table 14 - Distribution of babies with chromosomal defects according to maternal age

Maternal age (yrs)	Total number of babies with anomalies (Prevalence per 1000 births)					
	1993	1994	1995	1996	1997	93-97
<20	0 (n/a)	0 (n/a)	1 (7.0)	0 (n/a)	0 (n/a)	1 (1.0)
20-24	0 (n/a)	1 (1.0)	0 (n/a)	1 (1.0)	0 (n/a)	2 (0.4)
25-29	1 (0.5)	2 (1.2)	0 (n/a)	1 (0.6)	3 (1.8)	7 (0.8)
30-34	5 (3.6)	1 (0.7)	8 (6.2)	3 (2.3)	2 (1.7)	19 (2.9)
35-39	7 (11.4)	3 (5.2)	4 (7.2)	3 (5.2)	2 (3.4)	19 (6.5)
40-44	4 (31.3)	1 (8.6)	1 (8.5)	4 (31.7)	2 (17.4)	12 (19.9)
45+	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)	0 (n/a)
Total	**17 (3.3)**	**8 (1.7)**	**14 (3.0)**	**12 (2.4)**	**9 (1.9)**	**60 (2.4)**

MONTH OF BIRTH

Table 15 - Distribution of babies registered with congenital anomalies according to month of birth

Month	Total number of babies with anomalies (Prevalence per 1000 births)					
	1993	1994	1995	1996	1997	93-97
January	7 (17.1)	19 (43.2)	17 (43.1)	14 (32.3)	16 (40.6)	73 (35.2)
February	14 (37.1)	13 (34.4)	9 (26.1)	6 (15.5)	14 (38.0)	56 (30.2)
March	17 (39.0)	8 (19.6)	9 (23.8)	13 (31.7)	24 (60.6)	71 (35.0)
April	13 (31.5)	16 (41.7)	18 (47.5)	15 (38.0)	15 (39.2)	77 (39.4)
May	8 (18.3)	11 (29.2)	19 (49.0)	16 (42.7)	17 (48.3)	71 (36.7)
June	10 (25.1)	10 (24.9)	21 (55.1)	12 (28.2)	17 (38.9)	70 (34.2)
July	18 (38.5)	26 (56.5)	16 (37.4)	16 (34.2)	18 (36.9)	94 (40.6)
August	19 (42.6)	12 (29.0)	11 (33.1)	14 (33.5)	17 (43.0)	73 (36.4)
September	10 (22.1)	18 (42.5)	17 (40.9)	15 (35.1)	25 (61.0)	85 (39.9)
October	17 (35.8)	19 (50.1)	12 (27.5)	16 (38.1)	8 (20.0)	72 (34.1)
November	20 (46.8)	24 (60.9)	14 (38.6)	9 (23.7)	17 (42.9)	84 (42.8)
December	10 (23.2)	11 (27.2)	20 (50.9)	20 (45.5)	15 (36.1)	76 (36.4)
Total	**163 (31.5)**	**187 (38.5)**	**183 (39.5)**	**166 (33.3)**	**203 (42.0)**	**902 (36.8)**

GEOGRAPHICAL DISTRIBUTION

Table 16 - Distribution of babies with congenital anomaly according to locality of mother at time of birth

Locality	Total number of babies with anomalies (Prevalence per 1000 births)					
	1993	1994	1995	1996	1997	93-97
Gozo	**13 (31.4)**	**14 (37.1)**	**10 (29.1)**	**15 (45.9)**	**15 (49.0)**	**67 (37.9)**
North West	**72 (27.1)**	**93 (36.7)**	**106 (43.0)**	**97 (36.0)**	**106 (39.8)**	**474 (36.4)**
West	23 (48.9)	19 (39.3)	17 (40.4)	19 (35.8)	17 (30.9)	95 (38.7)
North	20 (25.3)	29 (37.2)	37 (50.3)	33 (44.9)	28 (35.4)	147 (38.3)
Central	8 (28.3)	10 (41.0)	15 (54.9)	11 (37.1)	15 (49.3)	59 (42.1)
Central North	11 (18.9)	17 (32.3)	20 (37.2)	18 (30.7)	30 (54.6)	96 (34.5)
Central West	10 (18.8)	18 (36.1)	17 (34.0)	16 (29.4)	16 (34.3)	77 (30.3)
South East	**78 (37.1)**	**80 (40.9)**	**67 (36.8)**	**54 (27.6)**	**82 (43.3)**	**361 (37.1)**
Central East	14 (40.5)	17 (45.9)	12 (36.9)	12 (30.4)	26 (70.3)	81 (44.9)
Harbour	14 (46.1)	10 (43.1)	6 (26.8)	7 (23.6)	9 (32.5)	46 (34.5)
Central South	10 (29.6)	11 (36.5)	13 (46.9)	7 (27.5)	7 (28.6)	48 (33.9)
South	20 (52.5)	16 (47.5)	12 (35.3)	9 (26.7)	17 (50.6)	74 (42.8)
East	20 (27.3)	26 (36.4)	24 (36.6)	19 (28.2)	23 (34.4)	112 (32.5)
Total	**163 (31.5)**	**187 (38.5)**	**183 (39.5)**	**166 (33.3)**	**203 (41.7)**	**902 (36.8)**

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
TOTAL BIRTHS (live and fetal deaths)			**5172**	**4863**	**4633**	**4978**	**4864**	**24510**	
ANOMALIES CLASSIFIED WITHIN THE ICD-9 CHAPTER ON CONGENITAL ANOMALIES (7400-7599)									
NEURAL TUBE DEFECTS (7400-7420)			**6**	**7**	**7**	**8**	**5**	**33**	**13.46**
7400	ANENCEPHALY/EXENCEPHALY	M	1	2	0	1	1	5	2.04
		F	0	1	1	1	0	3	1.22
7401	CRANIORACHISCHISIS (TOTAL OR PARTIAL)	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	0	1	0.41
7411	MENINGOCELE	M	0	0	0	0	0	0	0.00
		F	0	2	0	0	0	2	0.82
7412	MYELOMENINGOCELE	M	1	0	3	2	0	6	2.45
		F	0	0	1	1	2	4	1.63
7419	SPINABIFIDA	M	1	0	1	0	0	2	0.82
		F	1	1	1	0	0	3	1.22
7420	CEPHALOCELE	M	1	1	0	0	0	2	0.82
		F	1	0	0	2	2	5	2.04
OTHER ANOMALIES OF NERVOUS SYSTEM (7421-7429)			**8**	**13**	**9**	**8**	**8**	**46**	**18.77**
7421	MICROCEPHALY	M	1	1	0	1	0	3	1.22
		F	2	2	2	1	1	8	3.26
7422	REDUCTION DEFORMITIES OF BRAIN	M	2	1	1	3	1	8	3.26
		F	0	1	0	0	3	4	1.63
7423	CONGENITAL HYDROCEPHALY	M	1	4	3	0	0	8	3.26
		F	0	3	3	2	0	8	3.26
7424	OTHER SPECIFIED ANOMALIES OF BRAIN	M	2	0	0	0	1	3	1.22
		F	0	1	0	0	0	1	0.41
7425	OTHER ANOMALIES OF SPINAL CORD	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	0	1	0.41
7428	OTHER SPECIFIED ANOMALIES OF NERVOUS SYSTEM	M	0	0	0	0	0	0	0.00
		F	0	0	0	0	2	2	0.82

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
CONGENITAL ANOMALIES OF EYE, EAR, FACE AND NECK (7430-7449)			**9**	**15**	**13**	**14**	**12**	**63**	**25.70**
7430	ANOPHTHALMUS / CRYPTOPHTHALMOS	M	0	0	0	0	0	0	0.00
		F	0	1	0	0	1	2	0.82
7431	MICROPHTHALMOS / APLASIA / DYSPLASIA / HYPOPLASIA OF EYE	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	0	1	0.41
7432	BUPHTHALMOS	M	1	0	0	0	0	1	0.41
		F	0	0	1	0	0	1	0.41
7434	COLOBOMA AND OTHER ANOMALIES OF ANTERIOR SEGMENTS	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	1	2	0.82
7435	CONGENITAL ANOMALIES OF POSTERIOR SEGMENT OF EYE+B91	M	0	0	0	0	0	0	0.00
		F	0	0	0	0	1	1	0.41
7436	CONGENITAL ANOMALIES OF EYELIDS, LACRIMAL SYSTEM AND ORBIT	M	0	1	0	1	0	2	0.82
		F	1	0	1	0	1	3	1.22
7438	OTHER SPECIFIED ANOMALIES OF EYE	M	1	0	0	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
7441	ACCESSORY AURICLE	M	1	1	1	2	2	7	2.86
		F	0	0	0	0	0	0	0.00
7442	OTHER SPECIFIED ANOMALIES OF EAR	M	2	3	2	2	5	14	5.71
		F	0	1	5	2	0	8	3.26
		UN	0	1	0	0	0	1	0.41
7443	UNSPECIFIED ANOMALIES OF EAR	M	0	0	0	0	0	0	0.00
		F	0	1	0	0	0	1	0.41
7444	ANOMALIES OF FIRST BRANCHIAL ARCH / INCLUDING PREAURICULAR SINUS	M	1	1	0	2	0	4	1.63
		F	0	2	2	2	0	6	2.45
7445	WEBBING OF NECK	M	0	1	0	0	0	1	0.41
		F	1	0	0	0	0	1	0.41
7448	OTHER SPECIFIED ANOMALIES OF FACE AND NECK	M	0	1	0	0	0	1	0.41
		F	0	1	0	0	0	1	0.41
7449	UNSPECIFIED ANOMALIES OF FACE AND NECK	M	1	0	1	0	0	2	0.82
		F	0	0	0	1	1	2	0.82

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
ANOMALIES OF CARDIOVASCULAR SYSTEM (7450-7479)			**77**	**113**	**85**	**124**	**113**	**512**	**208.89**
7450	COMMON ARTERIAL TRUNCUS	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	0	1	0.41
7451	DISCORDANT VENTRICULOARTERIAL / ATRIOVENTRICULAR CONNECTION	M	2	1	1	1	2	7	2.86
		F	0	2	0	2	0	4	1.63
7452	TETRALOGY OF FALLOT	M	1	0	1	0	3	5	2.04
		F	1	0	0	1	1	3	1.22
7453	SINGLE VENTRICLE	M	0	0	0	0	1	1	0.41
		F	0	0	0	0	0	0	0.00
7454	VENTRICULAR SEPTAL DEFECT	M	12	9	17	15	16	69	28.15
		F	12	17	18	18	20	85	34.68
7455	ATRIAL SEPTAL DEFECT OSTIUM SECUNDUM TYPE	M	10	16	14	13	14	67	27.34
		F	11	23	11	29	30	104	42.43
7456	ENDOCARDIAL CUSHION DEFECT	M	0	1	1	0	1	3	1.22
		F	3	1	1	7	0	12	4.90
7460	ANOMALIES OF PULMONARY VALVE	M	4	7	5	3	3	22	8.98
		F	6	9	4	3	6	28	11.42
7461	ANOMALIES OF TRICUSPID VALVE	M	1	0	0	0	0	1	0.41
		F	0	0	0	1	0	1	0.41
7462	EBSTEIN'S ANOMALY AND OTHER ANOMALIES OF TRICUSPID VALVE	M	0	0	1	0	1	2	0.82
		F	0	0	0	1	0	1	0.41
7463	AORTIC VALVE ATRESIA / STENOSIS	M	2	0	1	1	1	5	2.04
		F	0	0	1	0	0	1	0.41
7464	OTHER ANOMALIES OF AORTIC VALVE	M	1	1	0	0	0	2	0.82
		F	0	1	0	0	0	1	0.41
7465	MITRAL VALVE ATRESIA&STENOSIS	M	0	0	0	0	0	0	0.00
		F	0	1	0	0	0	1	0.41
7466	OTHER ANOMALIES OF MITRAL VALVE	M	0	0	0	0	0	0	0.00
		F	1	0	0	1	0	2	0.82
7467	HYPOPLASTIC LEFT HEART SYNDROME	M	1	0	2	0	0	3	1.22
		F	0	0	0	0	0	0	0.00

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
7468	OTHER MALFORMATIONS AND ANOMALIES OF THE HEART	M	0	1	1	3	0	5	2.04
		F	0	1	0	3	0	4	1.63
7470	PATENT DUCTUS ARTERIOSUS	M	2	6	1	4	4	17	6.94
		F	1	6	5	6	3	21	8.57
7471	COARCTATION OF AORTA	M	2	3	0	3	1	9	3.67
		F	0	3	0	1	2	6	2.45
7472	OTHER ANOMALIES OF THE AORTA	M	0	1	0	2	2	5	2.04
		F	0	0	0	0	0	0	0.00
7473	ANOMALIES OF PULMONARY ARTERY	M	0	0	0	0	2	2	0.82
		F	1	2	0	2	0	5	2.04
7474	ANOMALIES OF GREAT VEINS	M	1	0	0	0	0	1	0.41
		F	1	0	0	2	0	3	1.22
7479	UNSPECIFIED ANOMALIES OF CIRCULATORY SYSTEM	M	1	1	0	1	0	3	1.22
		F	0	0	0	0	0	0	0.00
ANOMALIES OF NOSE AND RESPIRATORY SYSTEM (7480-7489)			**5**	**11**	**4**	**3**	**8**	**31**	**12.65**
7480	CHOANAL ATRESIA / STENOSIS	M	0	1	0	0	3	4	1.63
		F	1	0	0	0	0	1	0.41
7481	OTHER ANOMALIES OF NOSE	M	0	0	0	0	0	0	0.00
		F	1	1	0	0	0	2	0.82
7483	OTHER ANOMALIES OF LARYNX, TRACHEA AND BRONCHUS+B198	M	0	1	1	1	1	4	1.63
		F	1	3	0	1	1	6	2.45
7485	AGENESIS, HYPOPLASIA AND DYSPLASIA OF LUNG	M	2	1	3	0	2	8	3.26
		F	0	2	0	1	1	4	1.63
		UN	0	1	0	0	0	1	0.41
7488	OTHER SPECIFIED ANOMALIES OF THE RESPIRATORY SYSTEM	M	0	1	0	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
CLEFT PALATE AND CLEFT LIP (7490-7493)			**13**	**12**	**12**	**9**	**11**	**57**	**23.26**
7490	CLEFT OF SECONDARY PALATE	M	5	3	3	4	3	18	7.34
		F	3	3	4	3	4	17	6.94
		UN	0	1	0	0	0	1	0.41

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
7491	CLEFT OF PRIMARY PALATE / CLEFT LIP	M	1	1	2	0	2	6	2.45
		F	1	0	0	0	0	1	0.41
7492	CLEFT OF PRIMARY AND SECONDARY PALATE	M	2	3	3	1	1	10	4.08
		F	1	1	0	1	1	4	1.63
ANOMALIES OF UPPER ALIMENTARY TRACT (7500-7509)			**7**	**14**	**4**	**3**	**15**	**43**	**17.54**
7500	ANKYLOGLOSSIA	M	0	0	1	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
7501	OTHER ANOMALIES OF THE TONGUE	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	0	1	0.41
7502	OTHER SPECIFIED ANOMALIES OF MOUTH AND PHARYNX	M	0	2	0	0	0	2	0.82
		F	2	1	0	0	1	4	1.63
7503	TRACHEO-OESOPHAGEAL FISTULA, OESOPHAGEAL ATRESIA AND STENOSIS	M	2	1	0	0	1	4	1.63
		F	0	1	0	0	0	1	0.41
7505	CONGENITAL HYPERTROPHIC PYLORIC STENOSIS	M	3	7	3	1	12	26	10.61
		F	0	2	0	1	1	4	1.63
OTHER ANOMALIES OF DIGESTIVE SYSTEM (7510-7519)			**4**	**2**	**9**	**9**	**4**	**28**	**11.42**
7511	ATRESIA AND STENOSIS OF SMALL INTESTINE	M	0	0	0	1	0	1	0.41
		F	0	0	1	0	1	2	0.82
7512	ATRESIA/STENOSIS OF LARGE INTESTINE, RECTUM AND ANAL CANAL	M	3	0	3	3	0	9	3.67
		F	0	0	0	1	0	1	0.41
		UN	0	0	0	0	1	1	0.41
7513	HIRSCHPRUNG'S DISEASE AND OTHER CONG. FUNCTIONAL DISORDERS OF COLON	M	0	1	2	3	0	6	2.45
		F	0	0	0	1	1	2	0.82
7514	ANOMALIES OF INTESTINAL FIXATION AND MESENTERIC ANOMALIES	M	0	0	0	0	0	0	0.00
		F	0	1	1	0	0	2	0.82
7515	OTHER ANOMALIES OF INTESTINE	M	1	0	0	0	0	1	0.41
		F	0	0	0	0	1	1	0.41
7516	ANOMALIES OF GALL BLADDER, BILE DUCTS AND LIVER	M	0	0	1	0	0	1	0.41
		F	0	0	1	0	0	1	0.41

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
ANOMALIES OF GENITAL ORGAN SYSTEM (7520-7529)			**19**	**14**	**14**	**12**	**17**	**76**	**31.01**
7520	ANOMALIES OF OVARIES	M	0	0	0	0	0	0	0.00
		F	0	2	0	0	0	2	0.82
7523	ANOMALIES OF UTERUS	M	0	0	0	0	0	0	0.00
		F	0	0	0	0	1	1	0.41
7524	ANOMALIES OF CERVIX, VAGINA AND EXTERNAL GENITALIA	M	0	0	0	0	0	0	0.00
		F	2	3	0	1	1	7	2.86
7525	ANOMALIES OF TESTICLES	M	1	3	4	4	1	13	5.30
		F	0	0	0	1	0	1	0.41
7526	HYPOSPADIAS / EPISPADIAS / CONGENITAL CHORDEE	M	12	2	9	4	13	40	16.32
		F	0	0	0	0	0	0	0.00
7527	INDETERMINATE SEX AND PSEUDOHERMAPHRODITISM	M	0	0	0	1	0	1	0.41
		F	2	0	0	0	0	2	0.82
		UN	0	1	0	0	1	2	0.82
7528	OTHER ANOMALIES OF MALE GENITAL TRACT	M	2	3	1	1	0	7	2.86
		F	0	0	0	0	0	0	0.00
ANOMALIES OF URINARY SYSTEM (7530-7539)			**18**	**24**	**14**	**17**	**15**	**88**	**35.90**
7530	RENAL AGENESIS AND DYSGENESIS	M	2	1	1	2	0	6	2.45
		F	0	0	0	0	1	1	0.41
7531	CYSTIC KIDNEY DISEASE	M	1	1	3	1	1	7	2.86
		F	1	2	0	0	1	4	1.63
7532	OBSTRUCTIVE DEFECTS OF RENAL PELVIS AND URETER	M	3	11	5	7	7	33	13.46
		F	5	4	1	0	3	13	5.30
		UN	0	2	0	0	0	2	0.82
7533	OTHER SPECIFIED ANOMALIES OF KIDNEY	M	1	2	1	2	1	7	2.86
		F	2	1	0	1	1	5	2.04
7534	OTHER SPECIFIED ANOMALIES OF URETER	M	1	0	0	1	0	2	0.82
		F	0	0	0	1	0	1	0.41
7536	ATRESIA & OTHER CONGENITAL STENOSIS OF URETHRA & BLADDER NECK	M	1	0	0	2	0	3	1.22
		F	0	0	1	0	0	1	0.41
7538	OTHER SPECIFIED ANOMALIES OF BLADDER AND URETHRA	M	1	0	2	0	0	3	1.22
		F	0	0	0	0	0	0	0.00

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
ANOMALIES OF MUSCULOSKELETAL SYSTEM (7540-7549)			**10**	**26**	**31**	**12**	**21**	**100**	**40.80**
7540	CONG. MUSCULOSKELETAL DEFORMITIES OF SKULL, FACE AND JAW	M	0	5	0	1	2	8	3.26
		F	0	0	1	0	2	3	1.22
7543	CONGENITAL DISLOCATION OF HIP	M	1	0	1	0	1	3	1.22
		F	1	7	5	1	1	15	6.12
7544	CONGENITAL GENU RECURVATUM AND BOWING OF LONG BONES OF LEG	M	1	0	2	0	0	3	1.22
		F	0	1	0	0	0	1	0.41
7545	VARUS DEFORMITIES OF FEET	M	2	2	9	3	5	21	8.57
		F	0	1	3	3	5	12	4.90
7546	VALGUS DEFORMITIES OF FEET	M	0	1	2	0	1	4	1.63
		F	0	0	0	0	0	0	0.00
7547	OTHER DEFORMITIES OF FEET	M	1	5	4	4	2	16	6.53
		F	2	3	4	0	2	11	4.49
7548	OTHER SPECIFIED DEFORMITIES OF THORAX AND LIMBS	M	1	1	0	0	0	2	0.82
		F	1	0	0	0	0	1	0.41
ANOMALIES OF LIMBS (7550-7559)			**25**	**25**	**26**	**16**	**18**	**110**	**44.88**
7550	POLYDACTYLY / POLYSYNDACTYLY HANDS AND FEET	M	7	4	7	1	5	24	9.79
		F	4	5	1	6	3	19	7.75
7551	SYNDACTYLY HANDS AND FEET	M	1	3	2	3	2	11	4.49
		F	4	0	3	0	2	9	3.67
7552	REDUCTION DEFORMITIES OF UPPER LIMBS	M	0	2	3	0	1	6	2.45
		F	1	1	0	0	0	2	0.82
7553	REDUCTION DEFORMITIES OF LOWER LIMBS	M	0	0	2	1	0	3	1.22
		F	1	0	0	0	0	1	0.41
7555	OTHER ANOMALIES OF UPPER LIMB, INCLUDING SHOULDER GIRDLE	M	1	3	1	1	1	7	2.86
		F	1	2	2	0	1	6	2.45
7556	OTHER ANOMALIES OF LOWER LIMB, INCLUDING PELVIC GIRDLE	M	2	2	1	1	0	6	2.45
		F	0	1	1	2	1	5	2.04
7558	OTHER SPECIFIED ANOMALIES OF UNSPECIFIED LIMB	M	1	1	1	1	1	5	2.04
		F	2	0	2	0	0	4	1.63
7559	UNSPECIFIED ANOMALIES OF UNSPECIFIED LIMB	M	0	1	0	0	0	1	0.41
		F	0	0	0	0	1	1	0.41

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
OTHER MUSCULOSKELETAL ANOMALIES (7560-7569)			**9**	**9**	**9**	**14**	**11**	**52**	**21.22**
7560	ANOMALIES OF SKULL AND FACE BONES	M	2	1	1	1	1	6	2.45
		F	0	0	1	2	2	5	2.04
7561	ANOMALIES OF SPINE	M	0	0	0	0	1	1	0.41
		F	0	1	1	1	0	3	1.22
7563	OTHER ANOMALIES OF RIBS AND STERNUM	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	0	1	0.41
7564	CHONDRO / OSTEOCHONDRODYSTROPHY, CHONDRO / OSTEOCHONDRODYSPLASIA	M	0	0	0	0	0	0	0.00
		F	0	2	0	0	0	2	0.82
7565	OSTEODYSTROPHIES	M	0	0	0	0	0	0	0.00
		F	0	0	1	1	1	3	1.22
7566	ANOMALIES OF DIAPHRAGM	M	0	0	3	1	3	7	2.86
		F	2	3	0	3	0	8	3.26
7567	ANOMALIES OF ABDOMINAL WALL	M	3	0	0	3	1	7	2.86
		F	1	2	1	1	1	6	2.45
		UN	0	0	0	0	1	1	0.41
7568	OTHER SPECIFIED ANOMALIES OF MUSCLE, TENDON, FASCIA & CONNECTIVE TISSUE	M	0	0	1	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
7569	UNSPECIFIED ANOMALIES OF MUSCULOSKELETAL SYSTEM	M	1	0	0	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
CONGENITAL ANOMALIES OF INTEGUMENT (7570-7579)			**13**	**10**	**8**	**6**	**9**	**46**	**18.77**
7571	ICHTHYOSIS CONGENITA	M	1	0	0	0	1	2	0.82
		F	1	0	0	1	0	2	0.82
7572	DERMATOGLYPHIC ANOMALIES	M	1	2	0	0	0	3	1.22
		F	0	0	0	0	0	0	0.00
7573	OTHER SPECIFIED ANOMALIES OF SKIN	M	5	3	2	2	2	14	5.71
		F	4	4	5	2	6	21	8.57
7574	SPECIFIED ANOMALIES OF HAIR	M	0	1	0	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
7575	SPECIFIED ANOMALIES OF NAILS	M	1	0	0	0	0	1	0.41
		F	0	0	1	1	0	2	0.82

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
CHROMOSOMAL ANOMALIES (7580-7589)			**17**	**8**	**14**	**12**	**9**	**60**	**24.48**
7580	DOWN SYNDROME	M	4	5	5	2	2	18	7.34
		F	11	2	3	6	1	23	9.38
7582	EDWARD SYNDROME AND OTHER TOTAL TRISOMIES OF E GROUP	M	0	0	1	1	2	4	1.63
		F	0	1	1	0	2	4	1.63
7583	AUTOSOMAL DELETION SYNDROMES	M	1	0	1	0	0	2	0.82
		F	0	0	1	1	0	2	0.82
7584	BALANCED AUTOSOMAL TRANSLOCATIONS IN NORMAL INDIVIDUAL	M	0	0	1	0	0	1	0.41
		F	1	0	1	0	0	2	0.82
7585	OTHER CONDITIONS DUE TO AUTOSOMAL ANOMALIES	M	0	0	0	0	0	0	0.00
		F	0	0	0	2	0	2	0.82
7586	ABERRATIONS OF THE X CHROMOSOME IN THE ABSENCE OF A Y CHROMOSOME	M	0	0	0	0	0	0	0.00
		F	0	0	0	0	1	1	0.41
7589	CONDITIONS DUE TO ANOMALY OF UNSPECIFIED CHROMOSOMES	M	0	0	0	0	1	1	0.41
		F	0	0	0	0	0	0	0.00
OTHER AND UNSPECIFIED CONGENITAL ANOMALIES			**5**	**4**	**3**	**6**	**4**	**22**	**8.98**
7591	ANOMALIES OF ADRENAL GLAND	M	0	0	0	0	0	0	0.00
		F	0	0	0	1	0	1	0.41
7593	SITUS INVERSUS	M	0	1	0	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
7596	HAMARTOSIS NEC	M	0	0	0	0	0	0	0.00
		F	1	0	0	0	0	1	0.41
7598	SYNDROMES AND MULTIPLE CONGENITAL ANOMALIES NEC	M	2	2	2	2	1	9	3.67
		F	2	1	1	3	3	10	4.08
OTHER ANOMALIES NOT CLASSIFIED WITHIN THE ICD-9 CHAPTER ON CONGENITAL ANOMALIES									
NEOPLASMS (1400-2399)			**1**	**0**	**4**	**2**	**2**	**9**	**3.67**
1905	MALIGNANT NEOPLASM OF RETINA	M	1	0	1	0	0	2	0.82
		F	0	0	0	0	1	1	0.41

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
1940	MALIGNANT NEOPLASM OF SUPRARENAL GLAND	M	0	0	1	1	0	2	0.82
		F	0	0	1	0	0	1	0.41
1991	MALIGNANCY, UNSPECIFIED SITE	M	0	0	0	1	0	1	0.41
		F	0	0	0	0	0	0	0.00
2157	BENIGN NEOPLASM OF CONNECTIVE TISSUE TRUNK / BACK NOS	M	0	0	0	0	1	1	0.41
		F	0	0	0	0	0	0	0.00
2163	BENIGN NEOPLASM SKIN OF OTHER AND UNSPECIFIED PARTS OF FACE	M	0	0	0	0	0	0	0.00
		F	0	0	1	0	0	1	0.41
ENDOCRINE, NUTRITIONAL, METABOLIC & IMMUNITY DISORDERS (2400-2799)			**5**	**4**	**6**	**3**	**7**	**25**	**10.20**
243	CONGENITAL HYPOTHYROIDISM	M	0	0	0	1	0	1	0.41
		F	3	3	2	0	1	9	3.67
2532	PANHYPOPITUITARISM	M	0	0	0	0	1	1	0.41
		F	0	0	0	0	0	0	0.00
2552	CONGENITAL ADRENAL HYPERPLASIA	M	0	0	0	0	0	0	0.00
		F	1	0	0	0	0	1	0.41
2598	OTHER ENDOCRINE DISORDERS NEC	M	0	0	0	0	0	0	0.00
		F	0	1	0	0	0	1	0.41
2701	PHENYLKETUNURIA	M	0	0	2	0	1	3	1.22
		F	0	0	0	0	0	0	0.00
2754	DISORDERS OF CALCIUM METABOLISM	M	0	0	1	0	1	2	0.82
		F	0	0	0	0	0	0	0.00
2762	ACIDOSIS	M	0	0	0	0	0	0	0.00
		F	0	0	0	0	1	1	0.41
2774	DISORDERS OF BILIRUBIN EXCRETION	M	0	0	0	0	1	1	0.41
		F	0	0	0	0	0	0	0.00
2778	OTHER SPECIFIED DISORDERS OF METABOLISM	M	0	0	0	1	0	1	0.41
		F	0	0	1	1	0	2	0.82
2779	UNSPECIFIED DISORDERS OF METABOLISM	M	1	0	0	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
2790	DEFICIENCY OF HUMORAL IMMUNITY	M	0	0	0	0	0	0	0.00
		F	0	0	0	0	1	1	0.41

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
CONDITIONS OF BLOOD AND BLOOD FORMING ORGANS (2800-2899)			**0**	**0**	**1**	**0**	**1**	**2**	**0.82**
2820	HEREDITARY SPHEROCYTOSIS	M	0	0	0	0	1	1	0.41
		F	0	0	0	0	0	0	0.00
2822	ANAEMIA DUE TO DISORDERS OF GLUTATHIONE METABOLISM	M	0	0	1	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
CONDITIONS OF NERVOUS SYSTEM AND SENSE ORGANS (3200-3899)			**3**	**2**	**1**	**2**	**1**	**9**	**3.67**
3301	CEREBRAL LIPIDOSIS INCLUDING GANGLIOSIDOSIS	M	2	0	0	0	1	3	1.22
		F	0	0	0	0	0	0	0.00
3309	CEREBRAL DEGENERATION, UNSPECIFIED	M	0	0	0	0	0	0	0.00
		F	1	0	0	0	0	1	0.41
3350	WERDNIG HOFFMANN DISEASE/INFANTILE SPINAL MUSCULAR ATROPHY	M	0	0	0	0	0	0	0.00
		F	0	1	0	0	0	1	0.41
3351	SPINAL MUSCULAR ATROPHY	M	0	0	0	1	0	1	0.41
		F	0	0	0	1	0	1	0.41
3592	MYOTONIC DISORDERS	M	0	0	0	0	0	0	0.00
		F	0	1	0	0	0	1	0.41
3780	CONVERGENT CONCOMITANT STRABISMUS	M	0	0	1	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
CONDITIONS OF THE CIRCULATORY SYSTEM (3900-4599)			**1**	**0**	**2**	**2**	**3**	**8**	**3.26**
4251	HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY	M	0	0	0	2	2	4	1.63
		F	0	0	1	0	1	2	0.82
4253	ENDOCARDIAL FIBROELASTOSIS	M	0	0	0	0	0	0	0.00
		F	0	0	1	0	0	1	0.41
4270	PAROXYSMAL SUPRAVENTRICULAR TACHYCARDIA	M	1	0	0	0	0	1	0.41
		F	0	0	0	0	0	0	0.00
CONDITIONS OF THE DIGESTIVE SYSTEM (5200-5799)			**2**	**3**	**2**	**2**	**2**	**11**	**4.49**
5240	MAJOR ANOMALIES OF JAW SIZE	M	1	2	1	0	2	6	2.45
		F	1	0	0	2	0	3	1.22
550	INGUINAL HERNIA	M	0	1	0	0	0	1	0.41
		F	0	0	1	0	0	1	0.41

Table 17 - All anomalies registered in Malta from 1993-1997 (as at December 1999)

CODE	TEXT	SEX	1993	1994	1995	1996	1997	93-97	Prev./10000
CONDITIONS ORIGINATING IN THE PERINATAL PERIOD (7600-7799)			**0**	**0**	**2**	**0**	**1**	**3**	**1.22**
7710	CONGENITAL RUBELLA	M	0	0	1	0	0	1	0.41
		F	0	0	1	0	0	1	0.41
7786	CONGENITAL HYDROCELE	M	0	0	0	0	1	1	0.41
		F	0	0	0	0	0	0	0.00
GRAND TOTAL			**257**	**317**	**280**	**285**	**297**	**1436**	**585.88**

Table 18-Comparison of prevalence rates of congenital anomalies in total births and livebirths Malta (1993-97), EUROCAT Registries (1990-94).

EUROCAT Subgroups (Annex 3) / *ICD-9 code*	CONGENITAL ANOMALIES	Number Malta	Prevalence /10,000 total births Malta	EUROCAT registries	Number Malta	Prevalence /10,000 live births Malta	EUROCAT registries with preg. termination
01	Neural Tube defects*	33	**13.5**	(5.6 - 15.9)	28	**11.5**	(1.3 - 7.3)
02-04	Other anomalies of the Nervous System *	46	**18.8**	(3.9 - 19.8)	39	**16**	(4.6 - 12.9)
05	Anomalies of the eye	15	**6.1**	(2.7 - 11.8)	14	**5.7**	(2.4 - 11.4)
06	Anomalies of the ear	31	**12.6**	(2.1 - 12.5)	28	**11.5**	(2.0 - 11.5)
07-09	Congenital Heart Disease **	513	**209.3**	(23.5 - 101.5)	506	**207.7**	(22.1 - 93.7)
ICD-7467	*Hypoplasia left heart****	*3*	***1.2***	*(1.5 - 3.8)*	*2*	***0.8***	*(0.9 - 3.1)*
ICD-7450	*Common Truncus*	*1*	***0.4***	*(0.2 - 3.1)*	*1*	***0.4***	*(0.1 - 2.1)*
ICD-7451	*Transposition of great arteries*	*11*	***4.5***	*(1.0 - 6.0)*	*11*	***4.5***	*(0.9 - 5.4)*
10	Respiratory System defects (excluding nose)	24	**9.8**	NA	23	**9.4**	NA
ICD-7490	*Cleft palate**	*36*	***14.7***	*(2.5 - 9.4)*	*36*	***14.8***	*(2.1 - 7.9)*
ICD-7491/2	*Cleft lip with or without palate*	*21*	***8.6***	*(3.3 - 15.3)*	*20*	***8.2***	*(2.7 - 14.4)*
12-14 excl 7505	Anomalies of digestive system excl. pyloric stenosis	33	**13.5**	(3.6 - 24.3)	32	**13.1**	(2.8 - 18.8)
ICD-7505	*Pyloric stenosis*	*30*	***12.2***	*(0.6 - 47.8)*	*30*	***12.3***	*(1.2 - 47.5)*
15-16	Anomalies of external genitals	74	**30.2**	(7.5 - 32.7)	72	**29.6**	(7.1 - 31.1)
ICD-75260	*Hypospadias (excl. glandular /coronal)*	*29*	***11.8***	*(6.1 - 27.8)*	*29*	***11.9***	*(6.0 - 27.3)*
17-19	Anomalies of the internal urogenital system	91	**37.1**	(6.9 - 66.5)	88	**36.1**	(5.4 - 52.9)
20-23	Limb defects	196	**80**	(24.8 - 89.2)	185	**75.9**	(21.5 - 77.0)
24-25	Other musculoskeletal & connective tissue defects *	90	**36.7**	(6.8 - 34.1)	80	**32.8**	(6.2 - 26.7)
ICD-7566	*Anomalies of diaphragm**	*15*	***6.1***	*(1.6 - 5.8)*	*15*	***6.2***	*(1.3 - 4.3)*
ICD-75670	*Omphalocele*	*8*	***3.3***	*(0.8 - 3.9)*	*5*	***2.1***	*(0.6 - 2.3)*
ICD-75671	*Gastroschisis*	*3*	***1.2***	*(0.3 - 2.3)*	*3*	***1.2***	*(0.2 - 1.7)*
30-31	Anomalies of integument & skin	46	**18.8**	NA	46	**18.9**	NA
32	Down Syndrome*	41	**16.7**	(11.5 - 28.0)	41	**16.8**	(7.5 - 12.5)
33	Other chromosomal anomalies, excl. Down syndrome*	19	**7.8**	(6.0 - 24.6)	19	**7.8**	(3.3 - 7.4)
34-38	Other anomalies	65	**26.5**	NA	62	**25.4**	NA

* Live birth prevalence in Malta is higher than that of other EUROCAT registries

** Both total prevalence and livebirth prevalence in Malta are higher than that of other EUROCAT registries

*** Both total prevalence and livebirth prevalence in Malta are lower than that of other EUROCAT registries

PART FOUR -

SPECIFIC ANOMALY GROUPS

NEURAL TUBE DEFECTS (NTD)

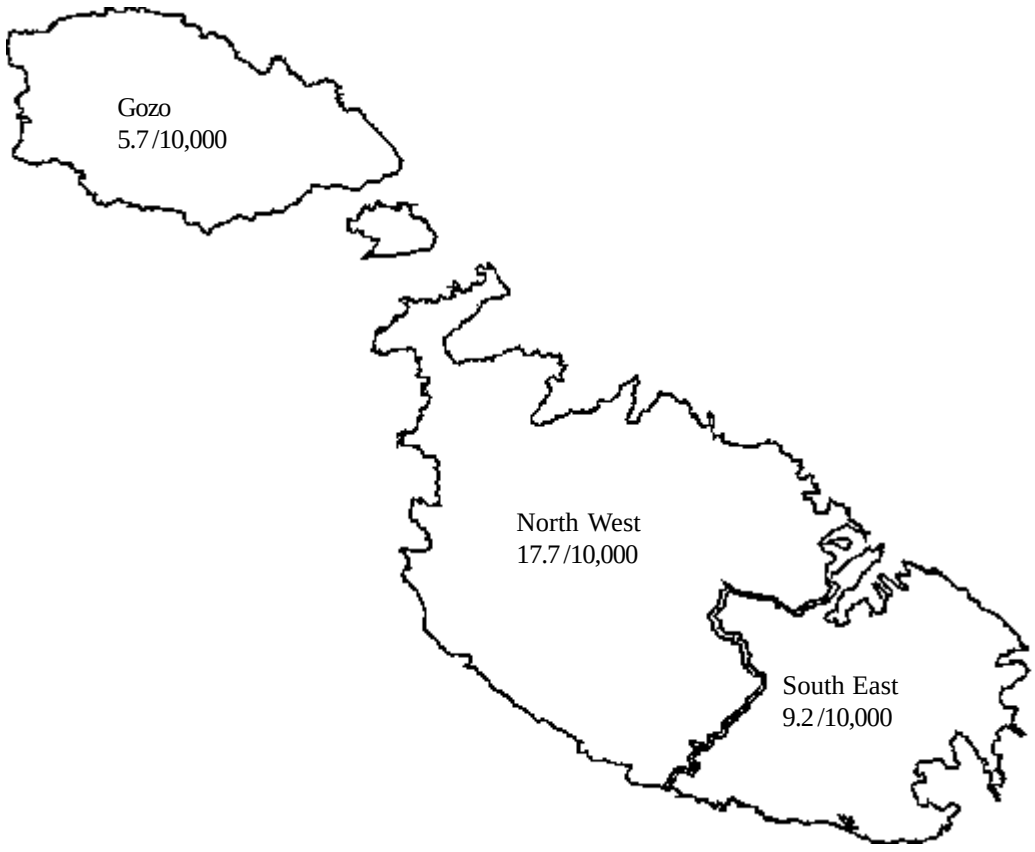
Neural tube defects include anencephaly, encephalocele, spina bifida and iniencephaly (ICD-9 codes 7400 - 7420)[1].

Geographical distribution

Table 19 - Distribution of NTD according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**18.8**	**15.8**	**24.3**	**22.3**	**7.5**	**17.7 (11.5 - 27.0)**
Anomalies	5	4	6	6	2	23
Total births	2656	2531	2467	2694	2662	13010
South East	**4.8**	**15.3**	**5.5**	**5.1**	**15.8**	**9.2 (4.5 - 18.2)**
Anomalies	1	3	1	1	3	9
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**30.6**	**0.0**	**5.7 (0.0 - 36.7)**
Anomalies	0	0	0	1	0	1
Total births	414	377	344	327	306	1768
Malta & Gozo	**11.6**	**14.4**	**15.1**	**16.1**	**10.3**	**13.5 (9.4 - 19.1)**

Figure 10 - Geographical distribution of NTD (1993-97)



The differences in prevalence do not reach statistical significance.

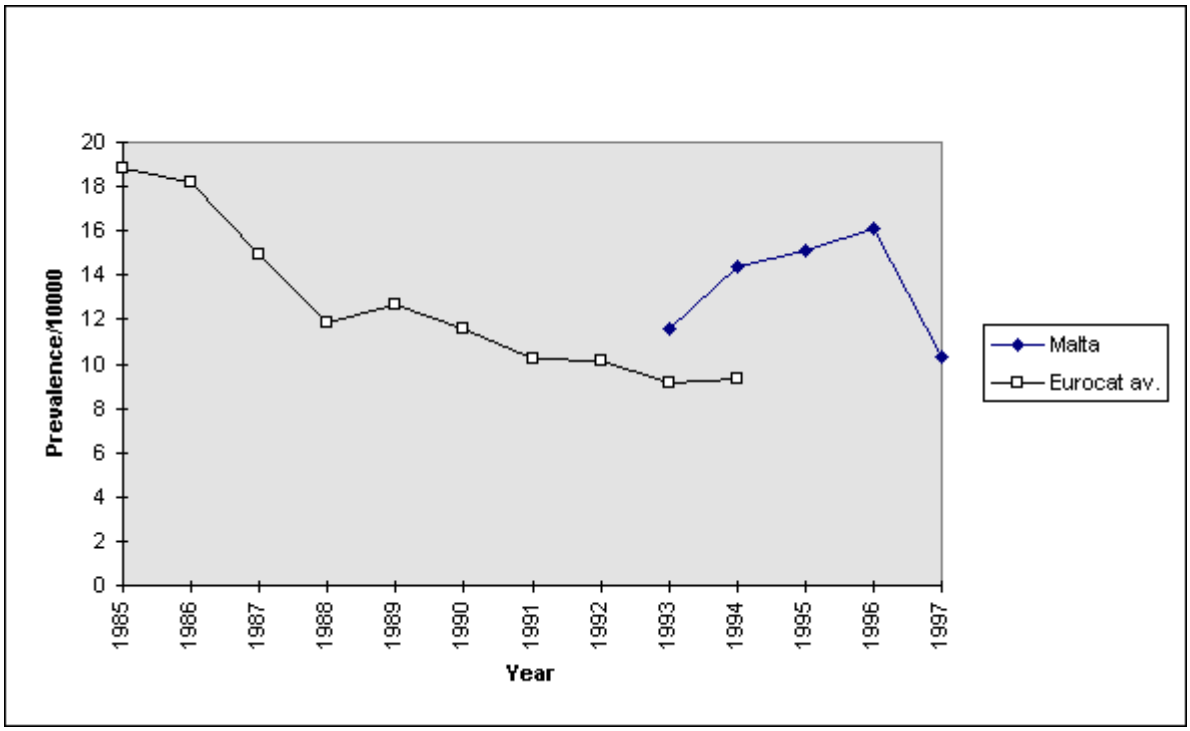
Time trends for NTD: Malta & EUROCAT

Table 20 - Time trend of total prevalence of NTDs Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	11.6	14.4	15.1	16.1	10.3	13.5 (1993-97)
EUROCAT average*	9.1	9.3				14.7 (1980-94)

*Data taken from EUROCAT Report 7 [1]

Figure 11 - Prevalence of NTD over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 13.5/10,000 total births (95% CI 9.4 to 19.1)
EUROCAT average 1990-1994 - 10.1/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 (5.6* to 15.9** /10,000 births)
* The lowest prevalence recorded was in the EUROCAT registry of - Switzerland
** The highest prevalence recorded was in the EUROCAT registry of - Glasgow

CLEFT LIP WITH OR WITHOUT CLEFT PALATE

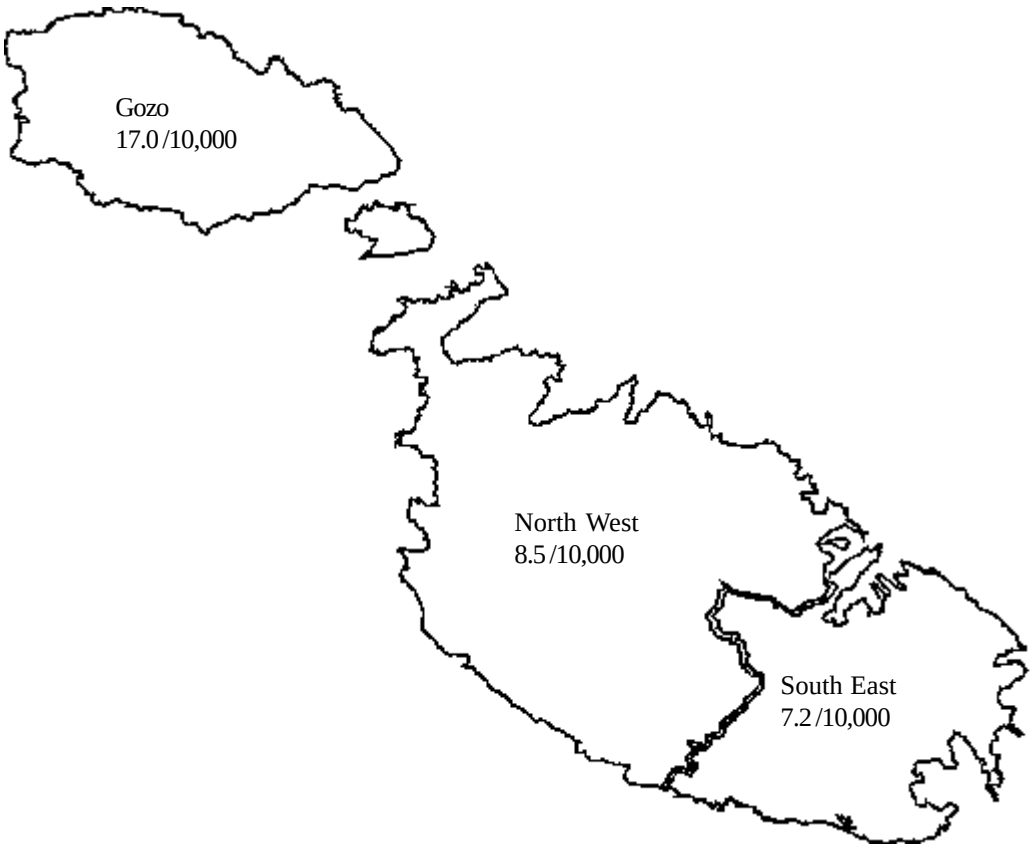
These include clefting of the upper lip with or without clefting of the maxillary alveolar process an hard and soft palate (ICD-9 codes: 7491 - 7492)[1].

Geographical distribution

Table 21 - Distribution of Cleft lip with or without cleft palate according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**3.8**	**15.8**	**16.2**	**3.7**	**3.7**	**8.5 (4.5 - 15.6)**
Anomalies	1	4	4	1	1	11
Total births	2656	2531	2467	2694	2662	13010
South East	**14.3**	**5.1**	**5.5**	**5.1**	**5.3**	**7.2 (3.2 - 15.5)**
Anomalies	3	1	1	1	1	7
Total births	2102	1955	1822	1957	1896	9732
Gozo	**24.2**	**0.0**	**0.0**	**0.0**	**65.4**	**17.0 (4.4 - 53.9)**
Anomalies	1	0	0	0	2	3
Total births	414	377	344	327	306	1768
Malta & Gozo	**9.7**	**10.3**	**10.8**	**4.0**	**8.2**	**8.6 (5.4 - 13.3)**

Figure 12 - Geographical distribution of Cleft lip with or without cleft palate (1993-97)



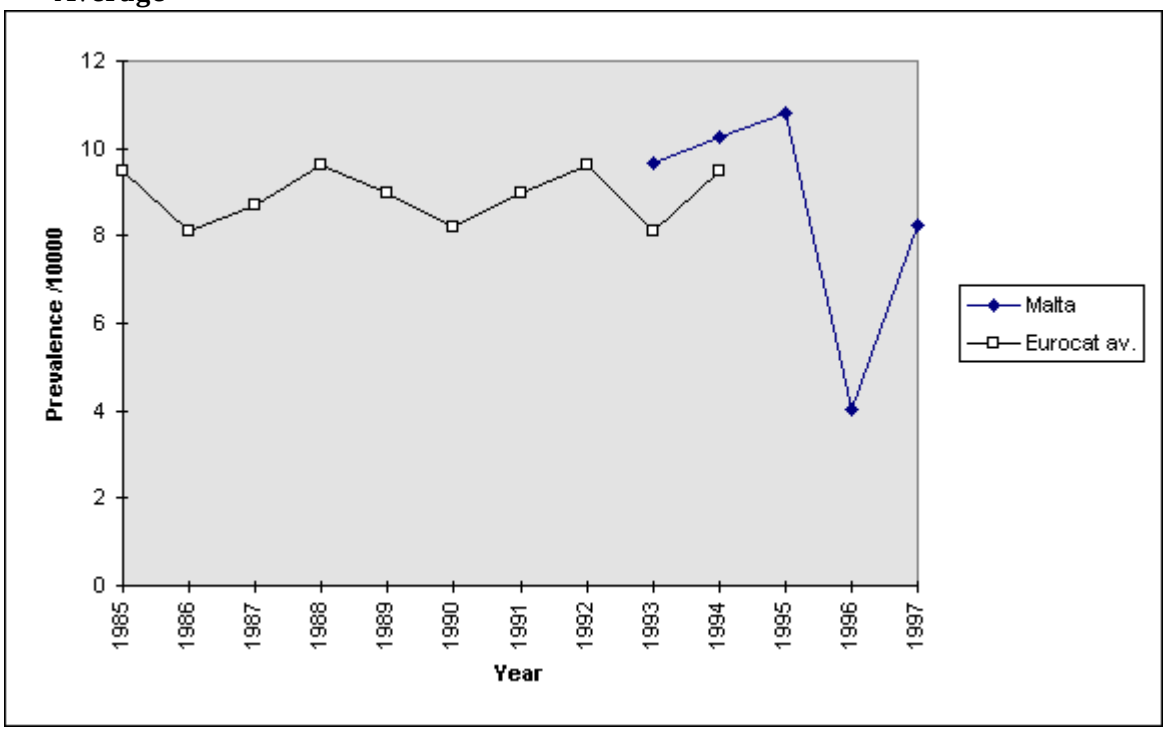
The difference in prevalence between the regions is not statistically significant.

Time trends for Cleft Lip with or without cleft palate: Malta & EUROCAT

Table 22 - Time trend of total prevalence of cleft lip Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	9.7	10.3	10.8	4.0	8.2	8.6 (1993-97)
EUROCAT average*	8.1	9.5	-	-	-	9.0 (1980-94)

Figure 13 - Prevalence of cleft lip with or without cleft palate over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 8.6/10,000 total births (95% CI 5.4 to 13.3)
EUROCAT average 1990-1994 - 8.9/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 3.3* to 15.3** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast
** The highest prevalence recorded was in the EUROCAT registry of - Northern Netherlands

CLEFT PALATE

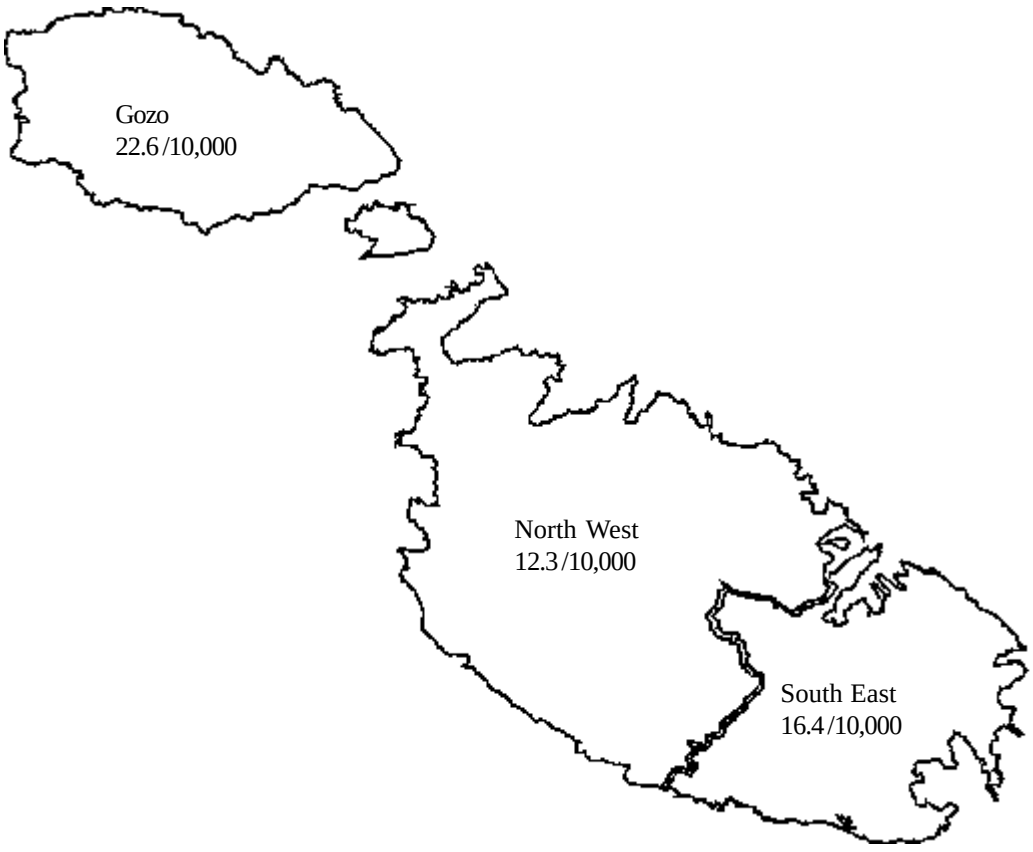
This includes fissure defects of the soft and / or hard palate(s) or submucous cleft without cleft lip (ICD-9 code: 7490)[1].

Geographical distribution

Table 23 - Distribution of Cleft Palate according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**11.3**	**4.0**	**20.3**	**7.4**	**18.8**	**12.3 (7.3 - 20.4)**
Anomalies	3	1	5	2	5	16
Total births	2656	2531	2467	2694	2662	13010
South East	**19.0**	**30.7**	**5.5**	**15.3**	**10.6**	**16.4 (9.7 - 27.3)**
Anomalies	4	6	1	3	2	16
Total births	2102	1955	1822	1957	1896	9732
Gozo	**24.2**	**0.0**	**29.1**	**61.2**	**0.0**	**22.6 (7.3 - 62.1)**
Anomalies	1	0	1	2	0	4
Total births	414	377	344	327	306	1768
Malta & Gozo	**15.5**	**14.4**	**15.1**	**14.1**	**14.9**	**14.7 (10.4 - 10.6)**

Figure 14 - Geographical distribution of Cleft Palate (1993-97)



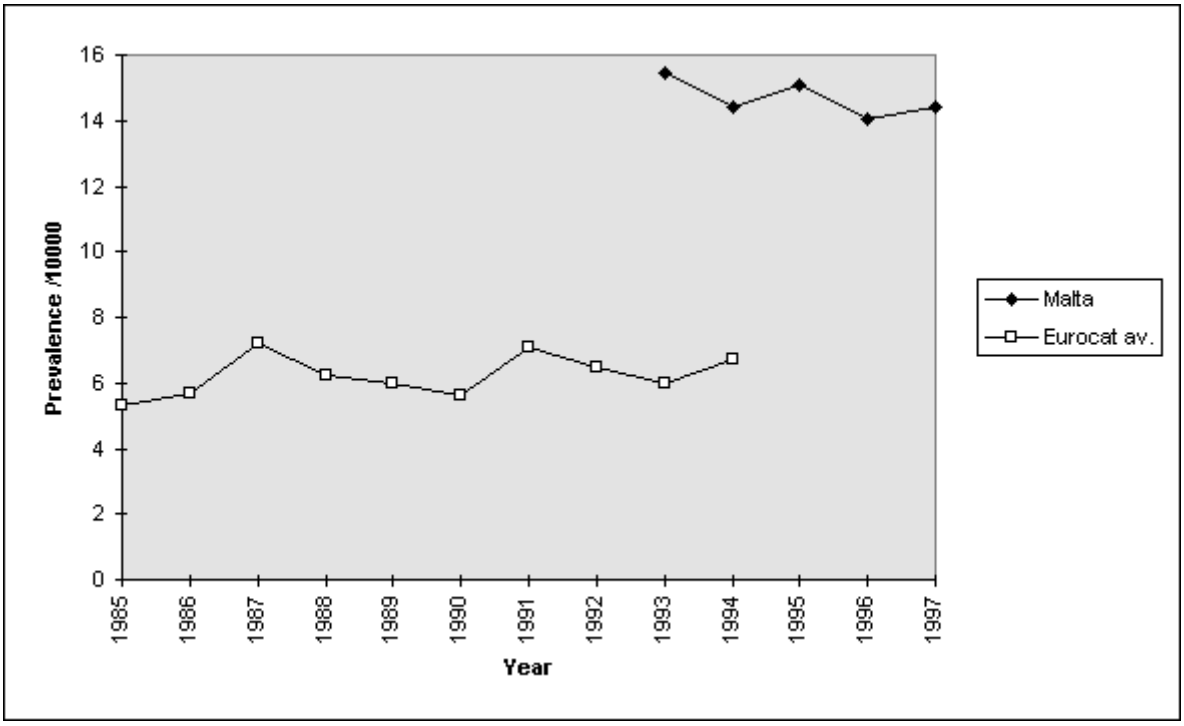
The difference in prevalence between the regions is not statistically significant.

Time trends for Cleft palate: Malta & EUROCAT

Table 24 - Time trend of total prevalence of cleft palate Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	15.5	14.4	15.1	14.1	14.9	14.7 (1993-97)
EUROCAT average*	6.0	6.7				6.5 (1980-94)

Figure 15 - Prevalence of cleft palate over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 14.7/10,000 total births (95% CI 10.4 to 20.6)
EUROCAT average 1990-1994 - 6.4/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 2.5* to 9.4** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast
** The highest prevalence recorded was in the EUROCAT registry of - Glasgow

Comments:
The prevalence of cleft palate in Malta is significantly higher than that reported by other European registries. All cases of cleft palate from 1993-97 in Malta were found as isolated defects.

CONGENITAL HEART DISEASE

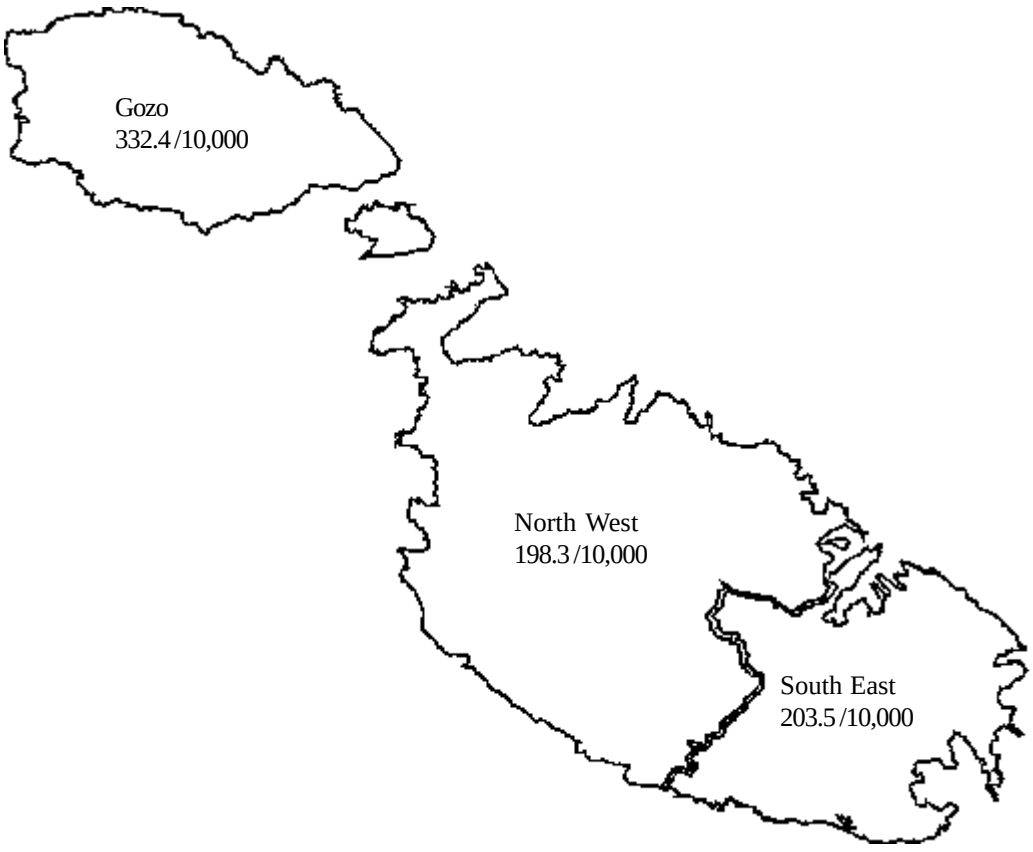
Congenital heart disease includes malformations of the heart, of great vessels, and endocardial fibroelastosis (ICD9 codes: 7450-7479 & 4253; EUROCAT Subgroups 07-09)[1].

Geographical distribution

Table 25 - Distribution of all congenital heart disease according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**113.0**	**225.2**	**174.3**	**274.7**	**202.9**	**198.3 (175- 224)**
Anomalies	30	57	43	74	54	258
Total births	2656	2531	2467	2694	2662	13010
South East	**180.8**	**230.2**	**181.1**	**178.8**	**247.9**	**203.5 (177 - 234)**
Anomalies	38	45	33	35	47	198
Total births	2102	1955	1822	1957	1896	9732
Gozo	**217.4**	**291.8**	**290.7**	**458.7**	**392.2**	**322.4 (247 - 419)**
Anomalies	9	11	10	15	12	57
Total births	414	377	344	327	306	1768
Malta & Gozo	**148.9**	**232.4**	**185.6**	**249.1**	**232.3**	**209.3 (194 - 231)**

Figure 16 - Geographical distribution of congenital heart disease (1993-97)



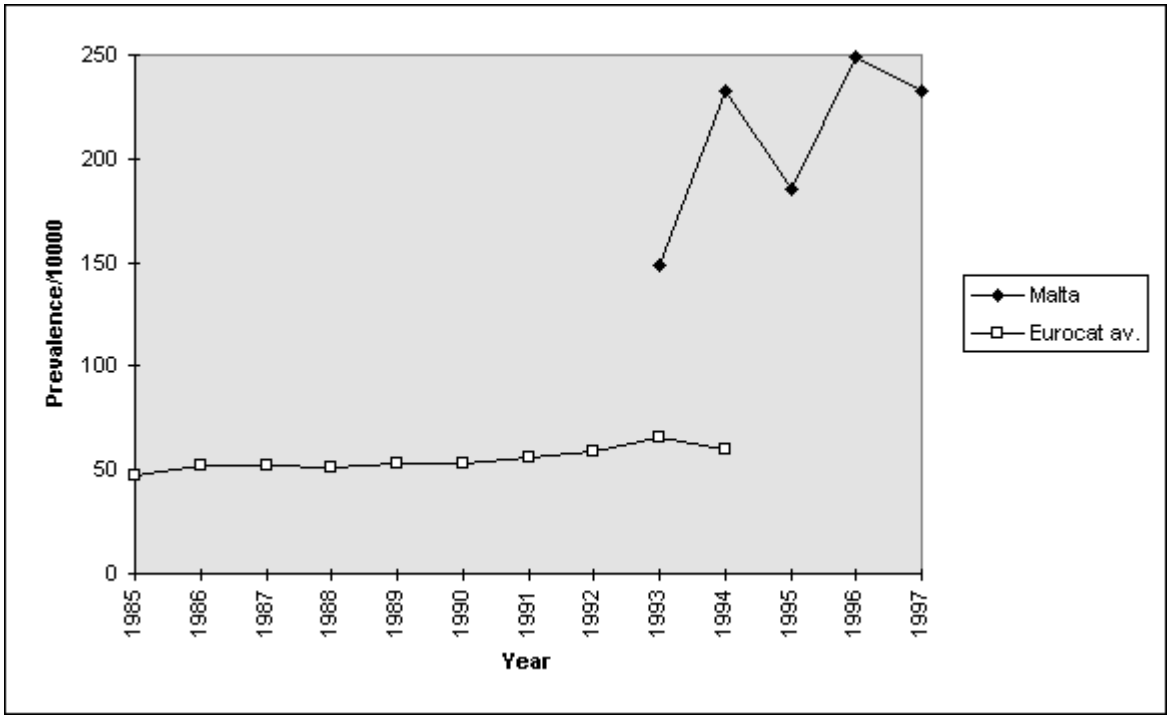
The difference in prevalence between Malta and Gozo is statistically significant with $p<0.01$.

Time trends for Congenital heart disease: Malta & EUROCAT

Table 26 - Time trend of total prevalence of Congenital heart disease Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	149	232	186	249	232	209 (1993-97)
EUROCAT average*	66	60				54 (1980-94)

Figure 17- Prevalence of Congenital heart disease over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 209/10,000 total births (95% CI 194 to 231)
EUROCAT average 1990-1994 - 59/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 23.5* to 101.5** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast
** The highest prevalence recorded was in the EUROCAT registry of - Strasbourg

ANOMALIES OF THE DIGESTIVE SYSTEM

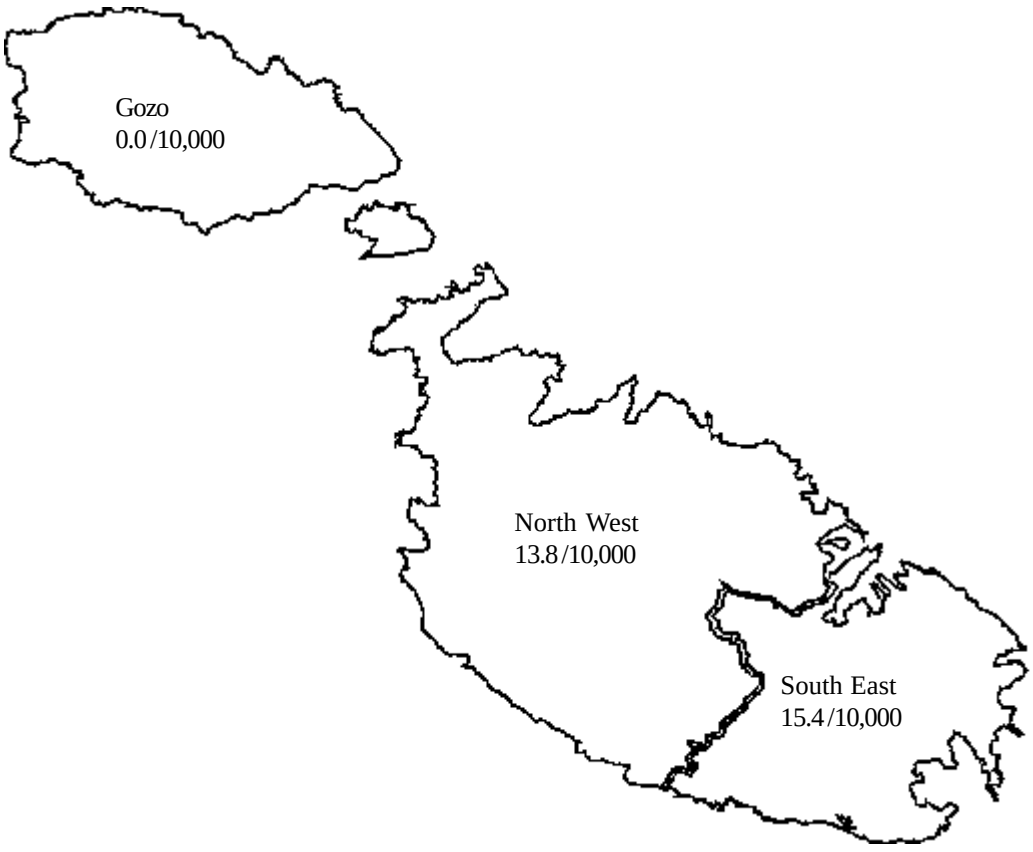
These include tracheo-oesophageal fistula, oesophageal atresia and stenosis , atresia and stenosis of small intestine, Meckel's diverticulum, colon disorders, anomalies of intestinal fixation and anomalies of gallbladder, bile ducts and liver , congenital pyloric stenosis excluded (EUROCAT subgroups 12, 13, 14 excluding pyloric stenosis)[1].

Geographical distribution

Table 27 - Distribution of defects of the digestive system according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**11.3**	**4.0**	**16.2**	**26.0**	**11.3**	**13.8 (8.5 - 22.3)**
Anomalies	3	1	4	7	3	18
Total births	2656	2531	2467	2694	2662	13010
South East	**14.3**	**15.3**	**27.4**	**10.2**	**10.6**	**15.4 (9.0 - 26.1)**
Anomalies	3	3	5	2	2	15
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**0.0**	**0.0**	**0.0 (0.0 - 27.0)**
Anomalies	0	0	0	0	0	0
Total births	414	377	344	327	306	1768
Malta & Gozo	**12.2**	**8.2**	**19.4**	**18.1**	**10.3**	**13.5 (9.4 - 19.1)**

Figure 18 - Geographical distribution of defects of the digestive system (1993-97)



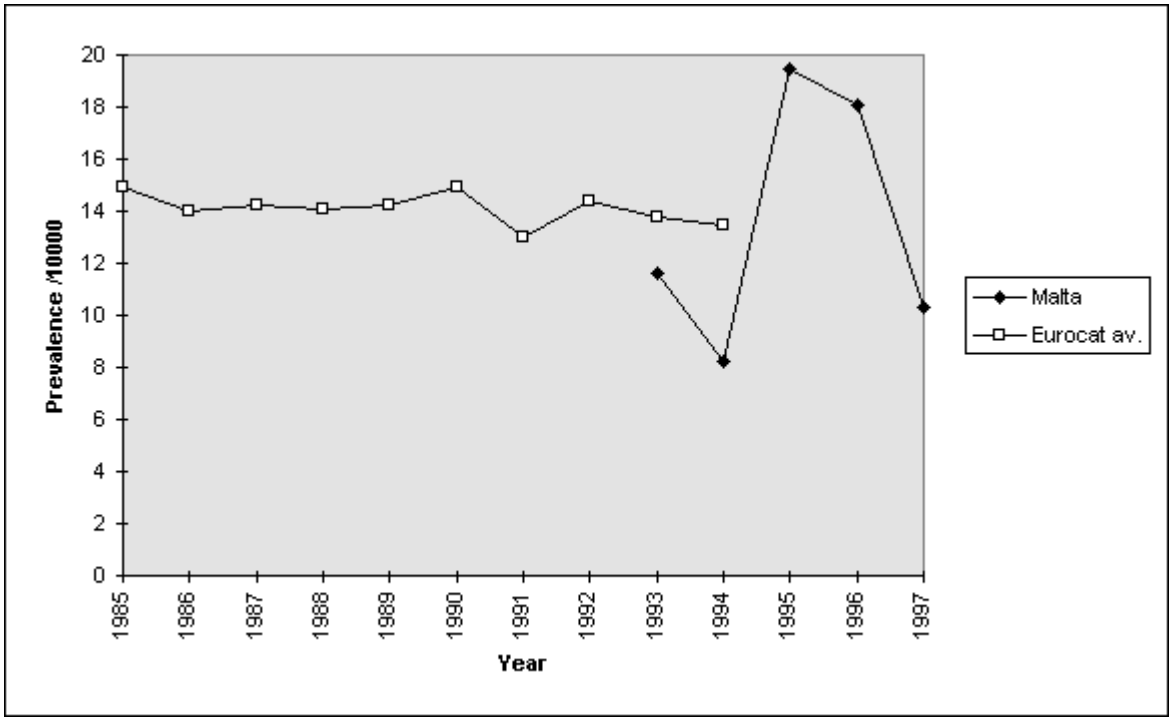
The differences in prevalence are not statistically significant.

Time trends for defects of the digestive system: Malta & EUROCAT

Table 28 - Time trend of total prevalence of defects of the digestive system Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	12.2	8.2	19.4	18.1	10.3	13.5(1993-97)
EUROCAT average*	13.8	13.5				14.3(1980-94)

Figure 19- Prevalence of defects of the digestive system over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 13.5/10,000 total births (95% CI 9.4 to 19.1)
EUROCAT average 1990-1994 - 13.9/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 3.6* to 24.3** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast
** The highest prevalence recorded was in the EUROCAT registry of - Glasgow

PYLORIC STENOSIS

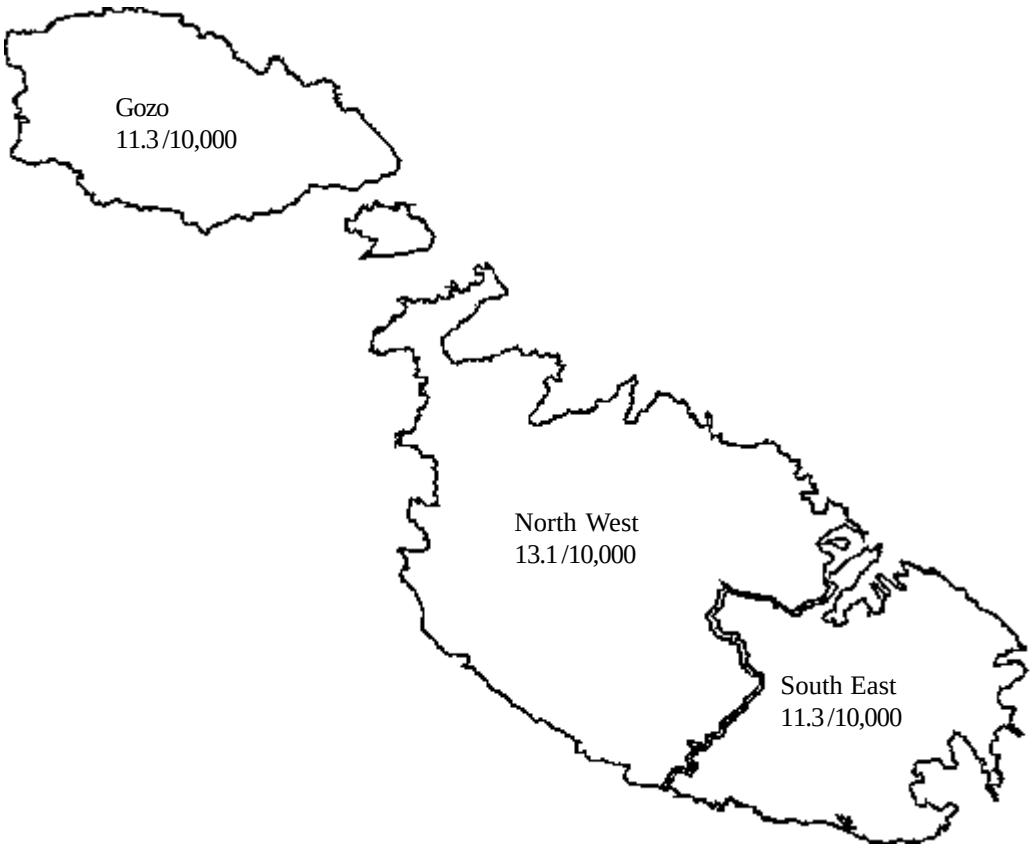
This is hypertrophy of muscular pylorus mainly detected after one month of life (ICD9 code: 7505)[1].

Geographical distribution

Table 29 - Distribution of congenital pyloric stenosis according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**0.0**	**15.8**	**8.1**	**3.7**	**37.6**	**13.1 (7.9 - 21.4)**
Anomalies	0	4	2	1	10	17
Total births	2656	2531	2467	2694	2662	13010
South East	**14.3**	**15.3**	**5.5**	**5.1**	**15.8**	**11.3 (5.9 - 20.9)**
Anomalies	3	3	1	1	3	11
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**53.1**	**0.0**	**0.0**	**0.0**	**11.3 (1.9 - 45.5)**
Anomalies	0	2	0	0	0	2
Total births	414	377	344	327	306	1768
Malta & Gozo	**5.8**	**18.5**	**6.5**	**4.0**	**26.7**	**12.2 (8.4 - 17.7)**

Figure 20 - Geographical distribution of pyloric stenosis (1993-97)



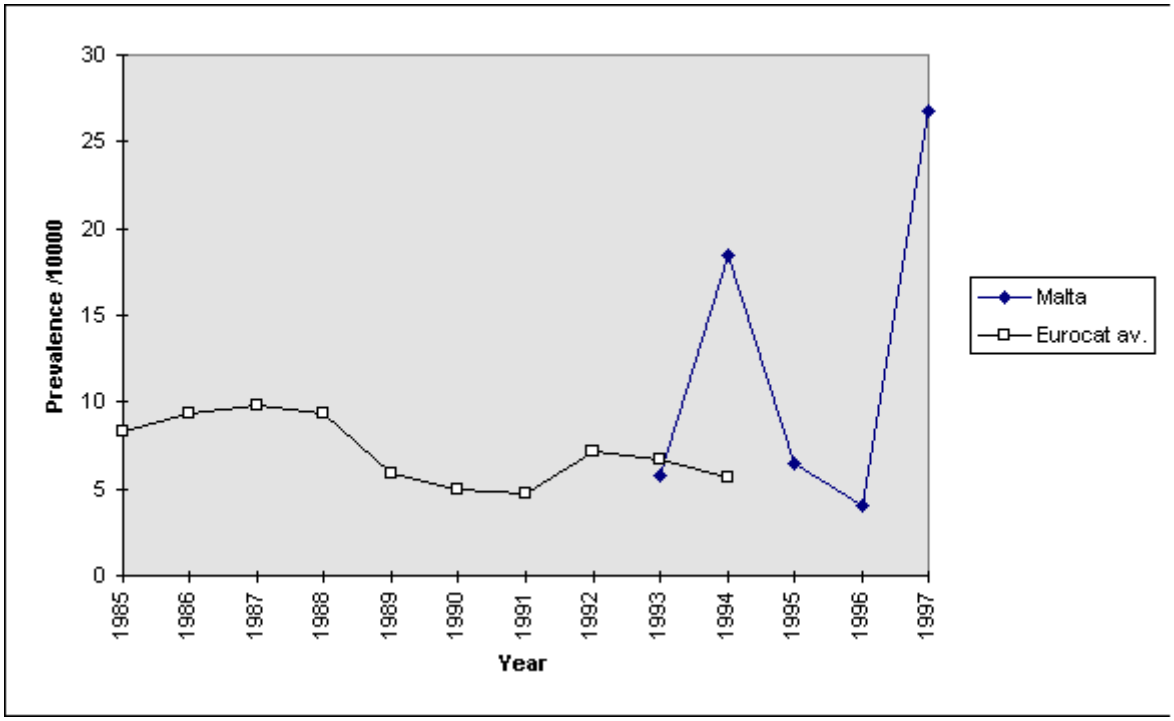
The differences in prevalence are not statistically significant.

Time trends for pyloric stenosis: Malta & EUROCAT

Table 30 - Time trend of total prevalence of congenital pyloric stenosis Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	5.8	18.5	6.5	4.0	26.7	12.2(1993-97)
EUROCAT average*	6.7	5.7				7.5(1980-94)

Figure 21- Prevalence of defects of pyloric stenosis over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 12.2/10,000 total births (95% CI 8.4 to 17.7)
EUROCAT average 1990-1994 - 5.9/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 0* to 47.8** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Odense, Paris, Hainaut-Namur
** The highest prevalence recorded was in the EUROCAT registry of - Antwerp

ANORECTAL ATRESIA AND STENOSIS

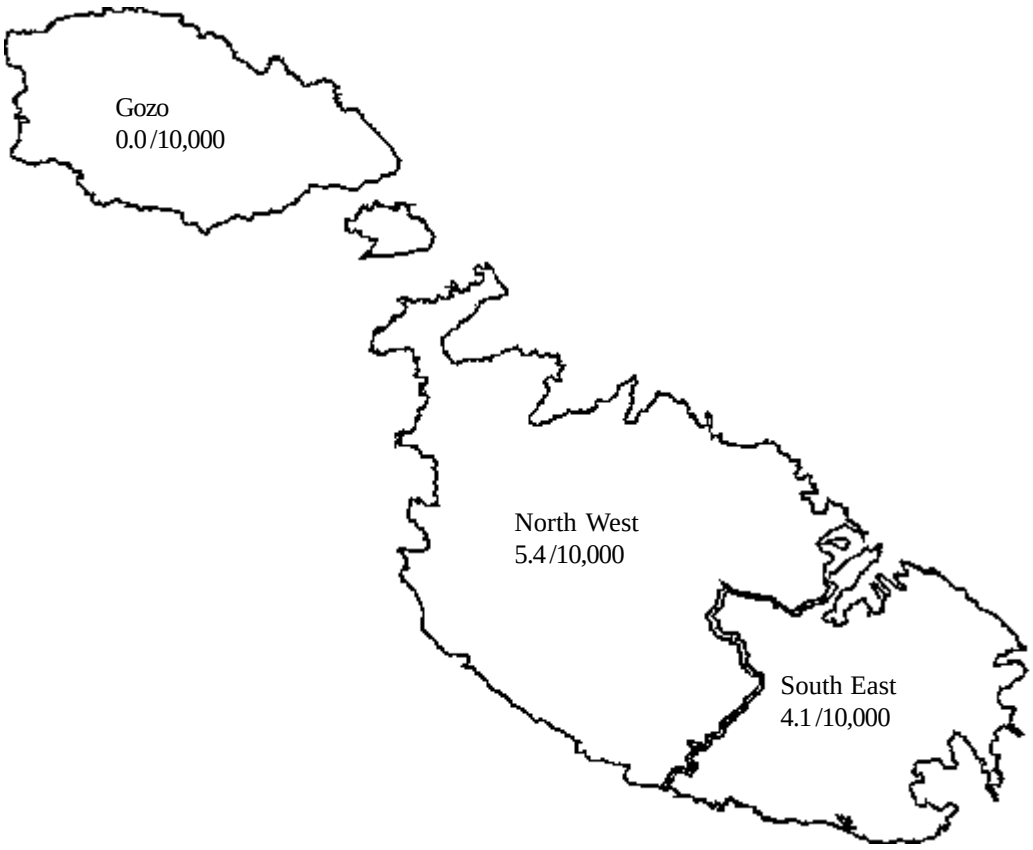
This includes imperforate anus or absence or narrowing of the communication canal between rectum and anus with or without fistula to neighbouring organs (ICD9 codes: 75121 - 75124)[1].

Geographical distribution

Table 31 - Distribution of anorectal atresia and stenosis according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**3.8**	**0.0**	**8.1**	**11.1**	**3.8**	**5.4 (2.4 - 11.6)**
Anomalies	1	0	2	3	1	7
Total births	2656	2531	2467	2694	2662	13010
South East	**9.5**	**0.0**	**5.5**	**5.1**	**0.0**	**4.1 (1.3 - 11.3)**
Anomalies	2	0	1	1	0	4
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**0.0**	**0.0**	**0.0 (0.0 - 27.0)**
Anomalies	0	0	0	0	0	0
Total births	414	377	344	327	306	1768
Malta & Gozo	**5.8**	**0.0**	**6.5**	**8.0**	**2.1**	**4.5 (2.4 - 8.3)**

Figure 22 - Geographical distribution of anorectal atresia and stenosis (1993-97)



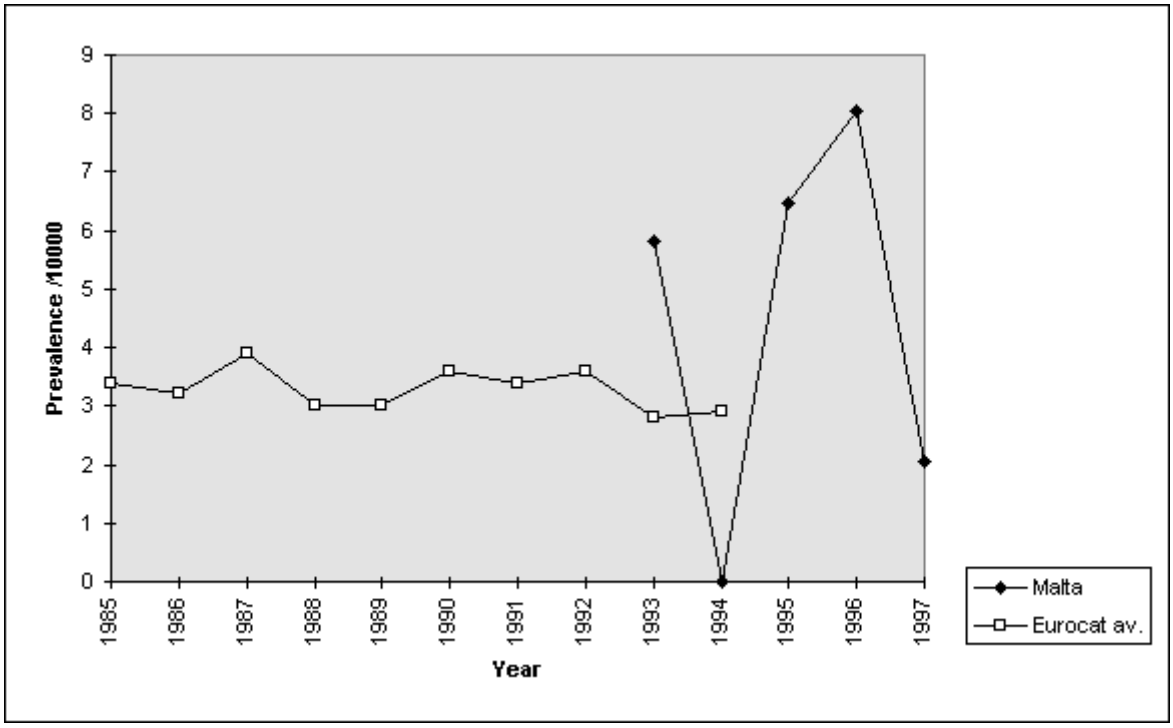
The differences in prevalence are not statistically significant.

Time trends for anorectal atresia and stenosis: Malta & EUROCAT

Table 32 - Time trend of total prevalence of anorectal atresia and stenosis Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	5.8	0.0	6.5	8.0	2.1	4.5(1993-97)
EUROCAT average*	2.8	2.9				3.4(1980-94)

Figure 23- Prevalence of defects of anorectal atresia and stenosis over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 4.5/10,000 total births (95% CI 2.4 to 8.3)
EUROCAT average 1990-1994 - 3.3/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 1.3* to 5.4** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast
** The highest prevalence recorded was in the EUROCAT registry of - Odense

ANOMALIES OF THE INTERNAL UROGENITAL SYSTEM

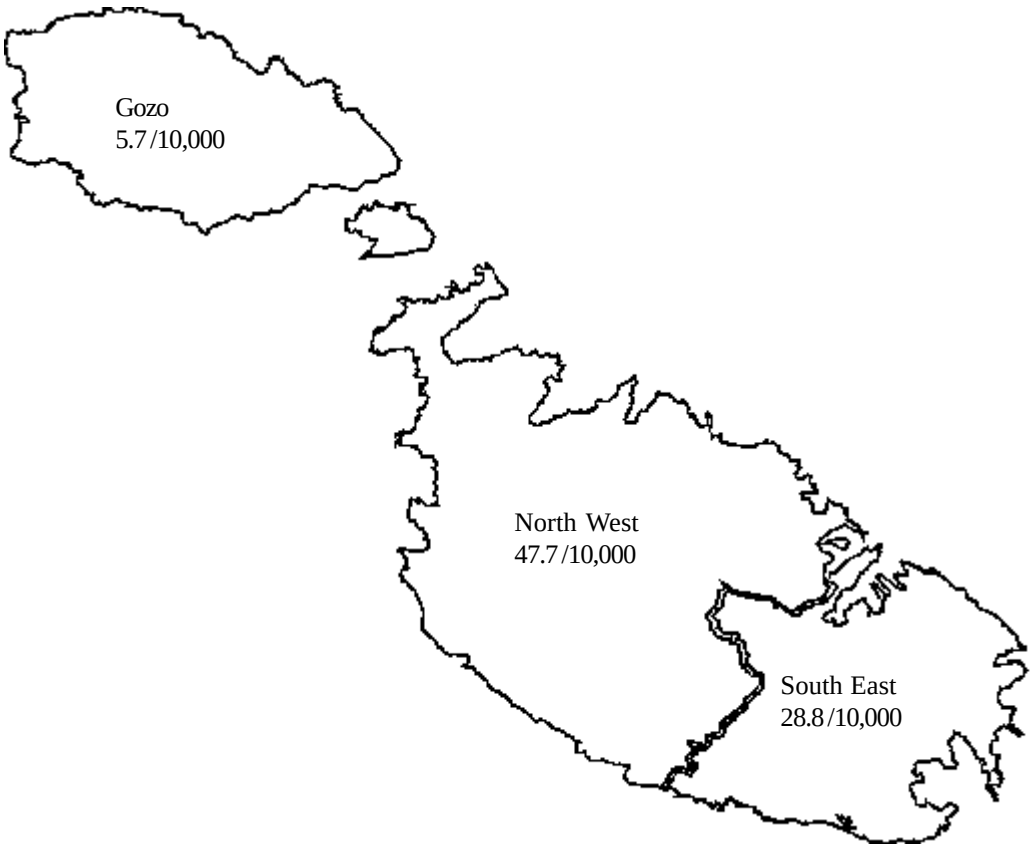
These include anomalies of ovaries, uterus and renal system (EUROCAT Subgroups 17-19)[1].

Geographical distribution

Table 33 - Distribution of defects of the internal urogenital system according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**67.8**	**71.1**	**20.3**	**44.5**	**33.8**	**47.7 (36.9 - 61.5)**
Anomalies	18	18	5	12	9	62
Total births	2656	2531	2467	2694	2662	13010
South East	**0.0**	**35.8**	**49.4**	**20.4**	**42.2**	**28.8 (19.5 - 42.2)**
Anomalies	0	7	9	4	8	28
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**30.6**	**0.0**	**5.7 (0.3 - 36.7)**
Anomalies	0	0	0	1	0	1
Total births	414	377	344	327	306	1768
Malta & Gozo	**34.8**	**51.4**	**30.2**	**34.2**	**35.0**	**37.1 (30.0 - 45.8)**

Figure 24 - Geographical distribution of defects of the internal urogenital system (1993-97)



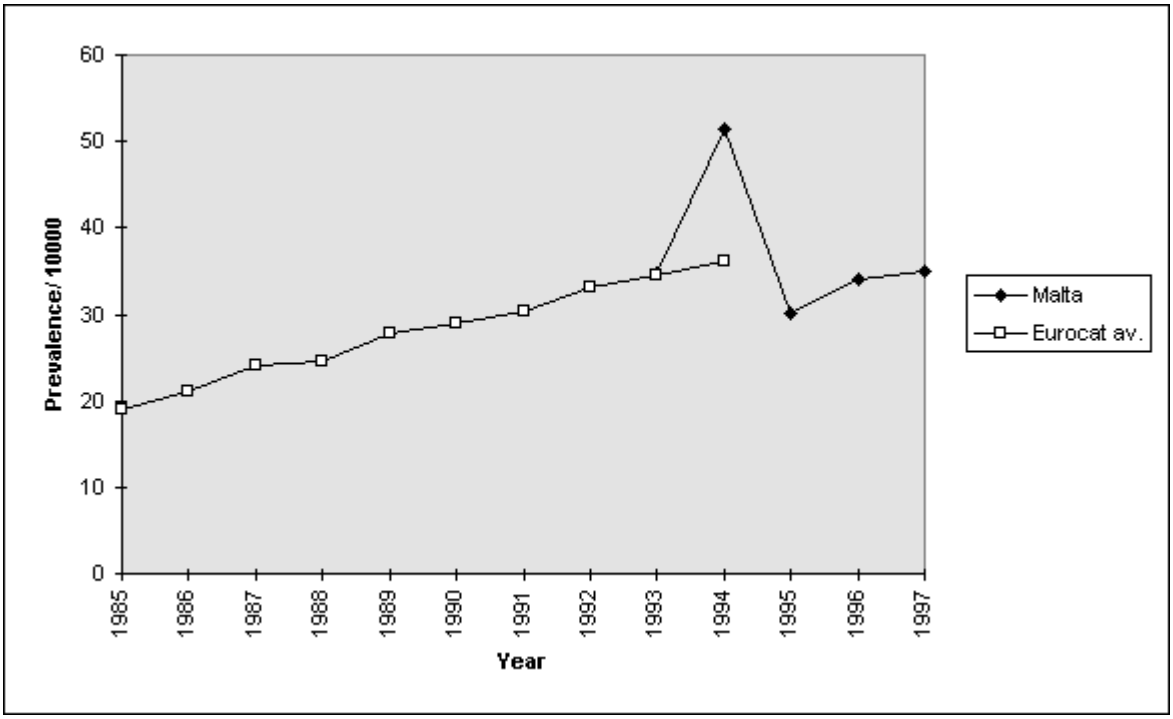
The differences in prevalence are statistically significant with $p<0.05$.

Time trends for defects of the internal urogenital system: Malta & EUROCAT

Table 34 - Time trend of total prevalence of defects of the internal urogenital system Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	34.8	51.4	30.2	34.2	35.0	37.1(1993-97)
EUROCAT average*	34.6	36.1				26.1(1980-94)

Figure 25- Prevalence of defects of the internal urogenital system over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 37.1/10,000 total births (95% CI 30.0 to 45.8)
EUROCAT average 1990-1994 - 32.6/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 6.9* to 66.5** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast
** The highest prevalence recorded was in the EUROCAT registry of - Paris

CYSTIC KIDNEY DISEASE

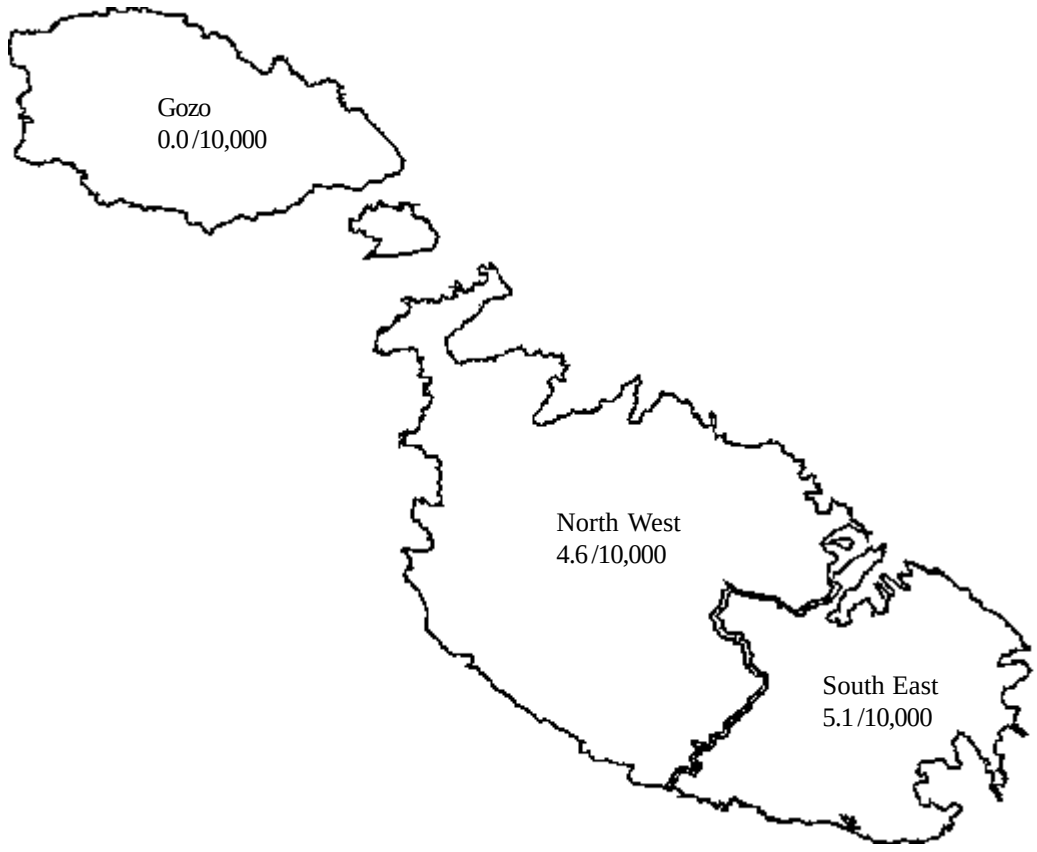
This includes presence of single or multiple cyst(s) infantile type, and enlarging kidney tissue (ICD9 code:7531)[1].

Geographical distribution

Table 35 - Distribution of cystic kidney defects according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**7.5**	**4.0**	**0.0**	**3.7**	**7.5**	**4.6 (1.9 - 10.6)**
Anomalies	2	1	0	1	2	6
Total births	2656	2531	2467	2694	2662	13010
South East	**0.0**	**10.2**	**16.5**	**0.0**	**0.0**	**5.1 (1.9 - 12.7)**
Anomalies	0	2	3	0	0	5
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**0.0**	**0.0**	**0.0 (0.0 - 27.0)**
Anomalies	0	0	0	0	0	0
Total births	414	377	344	327	306	1768
Malta & Gozo	**3.9**	**6.2**	**6.5**	**2.0**	**4.1**	**4.5 (2.3 - 8.3)**

Figure 26 - Geographical distribution of cystic kidney disease (1993-97)



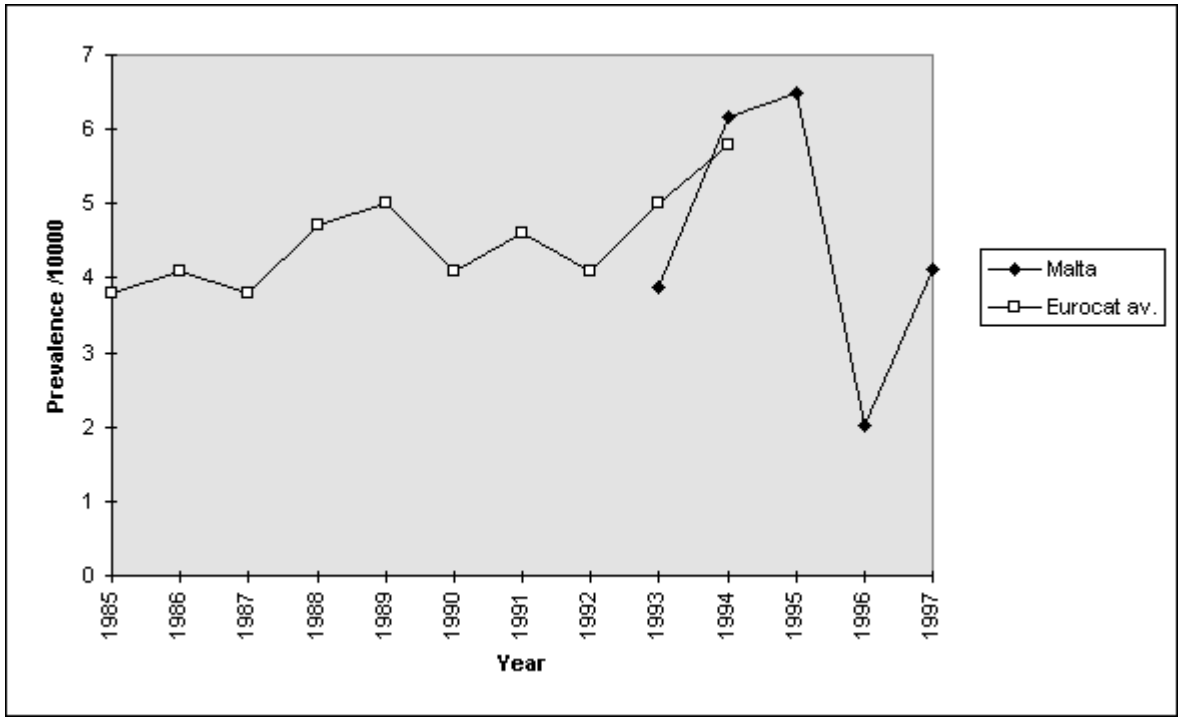
The differences in prevalence are not statistically significant.

Time trends for cystic kidney disease: Malta & EUROCAT

Table 36 - Time trend of total prevalence of cystic kidney disease: Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	3.9	6.2	6.5	2.0	4.1	4.5(1993-97)
EUROCAT average*	5.0	5.8				4.1(1980-94)

Figure 27- Prevalence of cystic kidney disease over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 4.5/10,000 total births (95% CI 2.3 to 8.3)
EUROCAT average 1990-1994 - 4.7/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 0.3* to 8.4** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Odense
** The highest prevalence recorded was in the EUROCAT registry of - Paris

LIMB REDUCTION DEFECTS

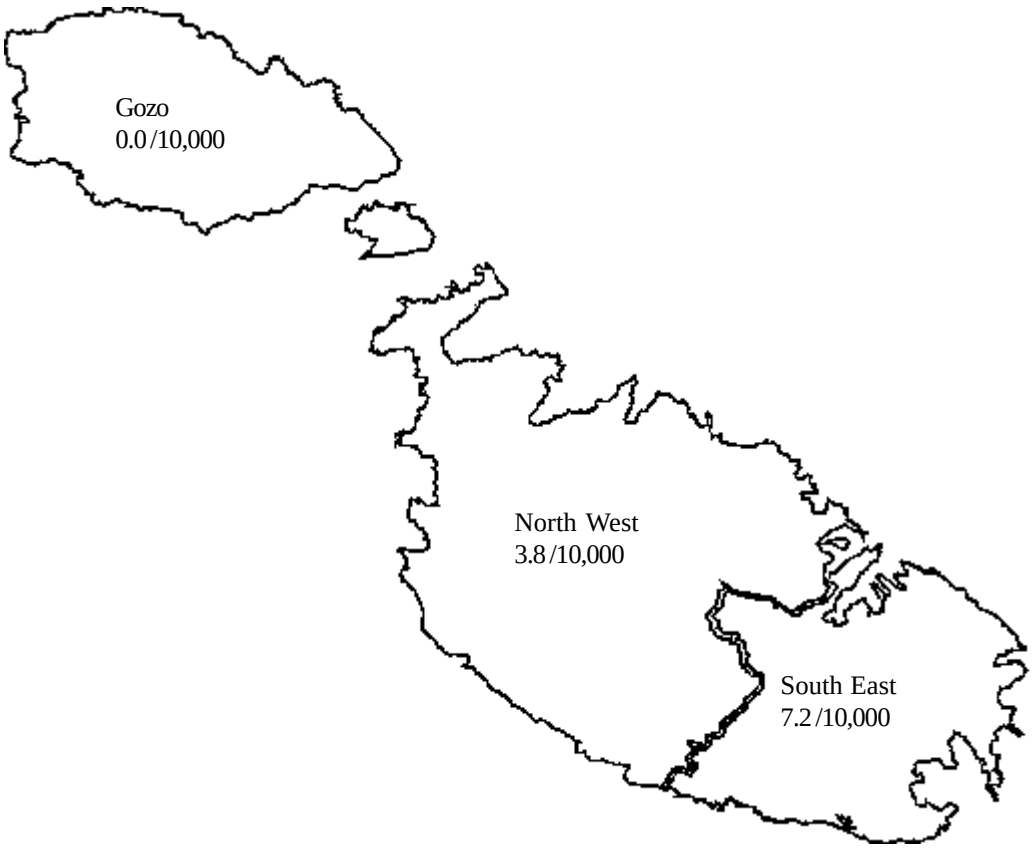
This includes total or partial absence or severe hypoplasia of skeletal structure of the limbs (ICD9 codes: 7552-7554)[1].

Geographical distribution

Table 37 - Distribution of limb reduction defects according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**3.8**	**7.9**	**8.1**	**0.0**	**0.0**	**3.8 (1.4 - 9.5)**
Anomalies	1	2	2	0	0	5
Total births	2656	2531	2467	2694	2662	13010
South East	**4.8**	**5.1**	**16.5**	**5.1**	**5.3**	**7.2 (3.2 - 15.5)**
Anomalies	1	1	3	1	1	7
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**0.0**	**0.0**	**0.0 (0.0 - 27.0)**
Anomalies	0	0	0	0	0	0
Total births	414	377	344	327	306	1768
Malta & Gozo	**3.9**	**6.2**	**10.8**	**2.0**	**2.1**	**4.9 (2.7 - 8.8)**

Figure 28 - Geographical distribution of limb reduction defects (1993-97)



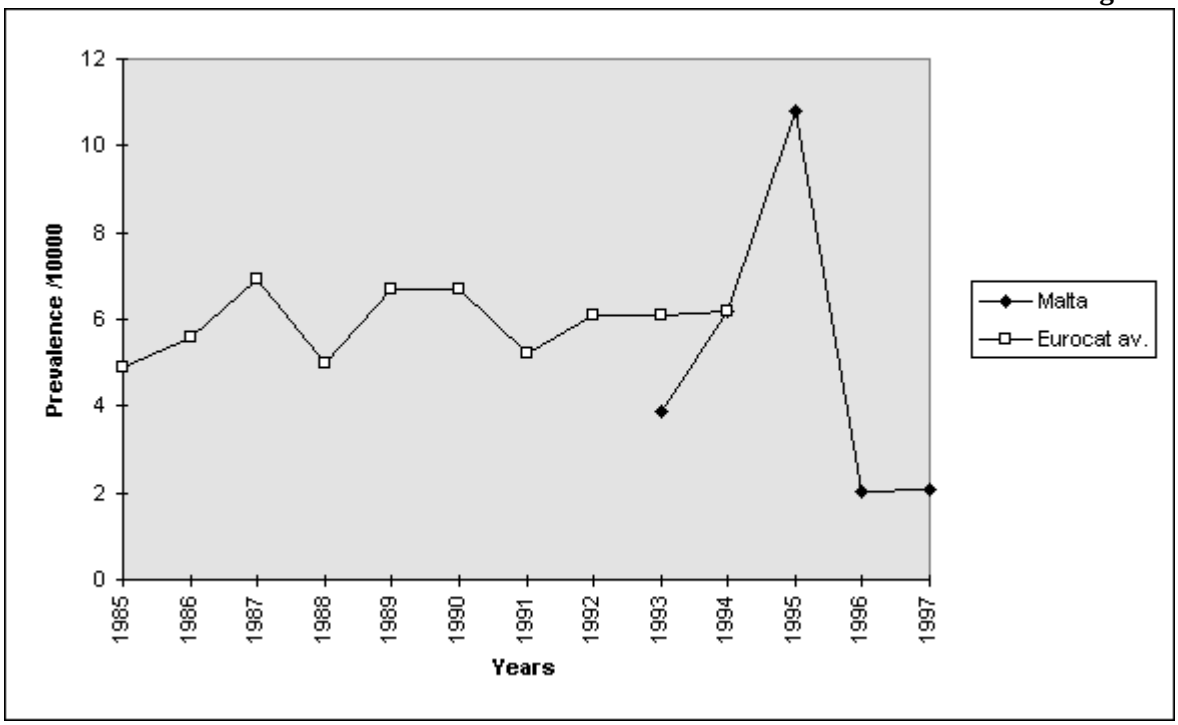
Differences in prevalence are not statistically significant.

Time trends for limb reduction defects: Malta & EUROCAT

Table 38 - Time trend of total prevalence of limb reduction defects: Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	3.9	6.2	10.8	2.0	2.1	4.9 (1993-97
EUROCAT average*	6.1	6.2				6.0 (1980-94

Figure 29- Prevalence of limb reduction defects over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 4.9/10,000 total births (95% CI 2.7 to 8.8)
EUROCAT average 1990-1994 - 6.1/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 1.5* to 9.8** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Galway
** The highest prevalence recorded was in the EUROCAT registry of - Asturias

POLYDACTYLY

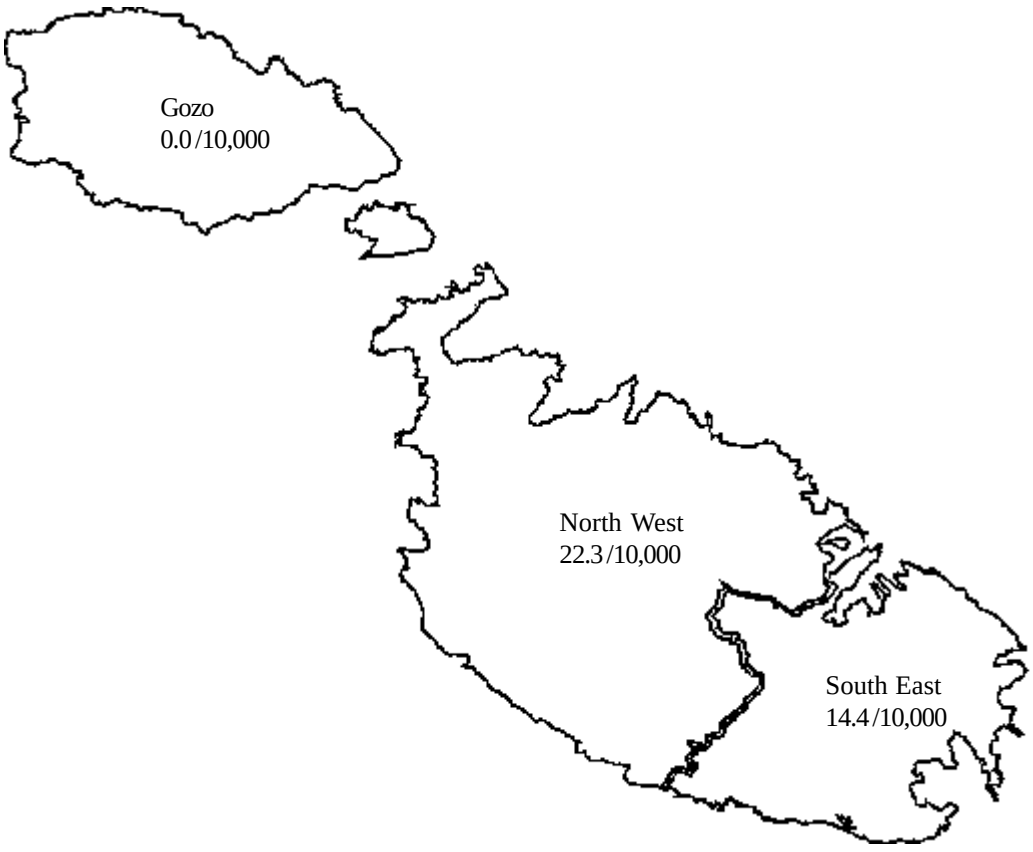
Partial or total extra digit on the radial (pre-axial), cubital (post axial) side of the hand or axial location, between 2nd, 3rd or 4th finger (ICD9 code: 7550)[1].

Geographical distribution

Table 39 - Distribution of polydactyly according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**30.1**	**19.8**	**20.3**	**18.6**	**22.5**	**22.3 (15.2 - 32.5)**
Anomalies	8	5	5	5	6	29
Total births	2656	2531	2467	2694	2662	13010
South East	**14.3**	**20.5**	**16.5**	**10.2**	**10.6**	**14.4 (8.2 - 24.8)**
Anomalies	3	4	3	2	2	14
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**0.0**	**0.0**	**0.0 (0.0 - 27.0)**
Anomalies	0	0	0	0	0	0
Total births	414	377	344	327	306	1768
Malta & Gozo	**21.3**	**18.5**	**17.3**	**14.1**	**16.5**	**17.5 (12.9 - 23.9)**

Figure 30 - Geographical distribution of polydactyly (1993-97)



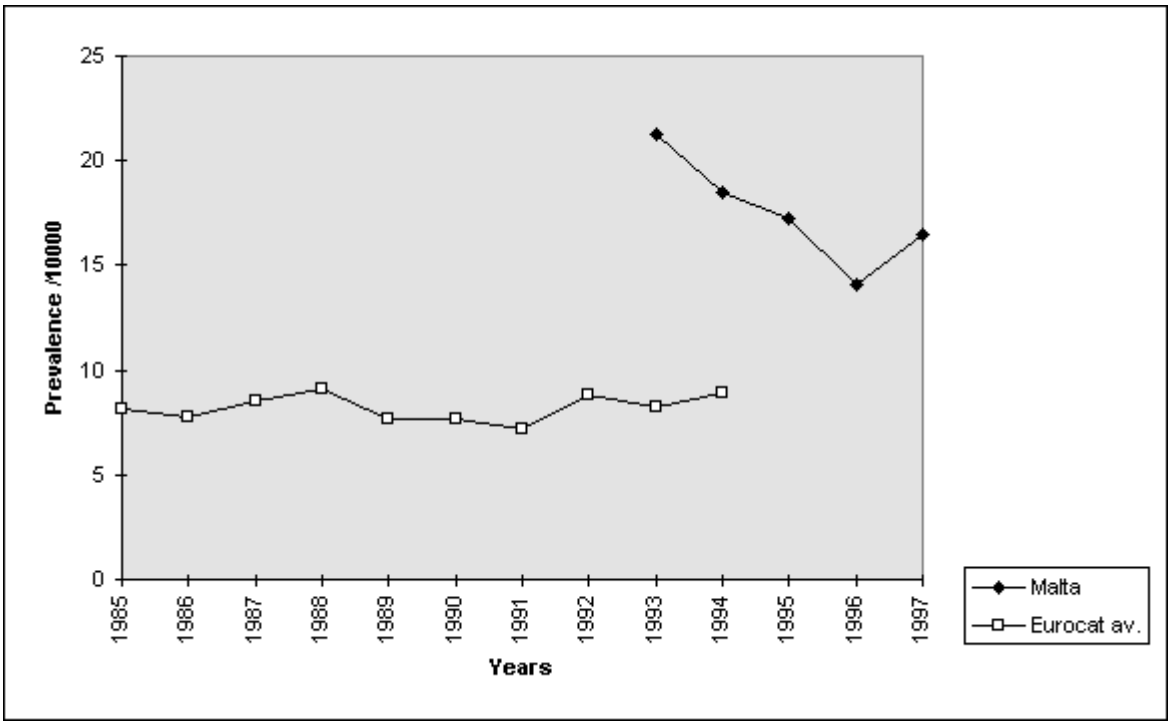
The differences in prevalence are not statistically significant.

Time trends for polydactyly: Malta & EUROCAT

Table 40 - Time trend of total prevalence of polydactyly: Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	21.3	18.5	17.3	14.1	16.5	17.5 (1993-97)
EUROCAT average*	8.2	8.9				8.3 (1980-94)

Figure 31- Prevalence of polydactyly over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 17.5/10,000 total births (95% CI 12.9 to 23.9)
EUROCAT average 1990-1994 - 8.2/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 4.1* to 18.0** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast
** The highest prevalence recorded was in the EUROCAT registry of - Paris

SYNDACTYLY

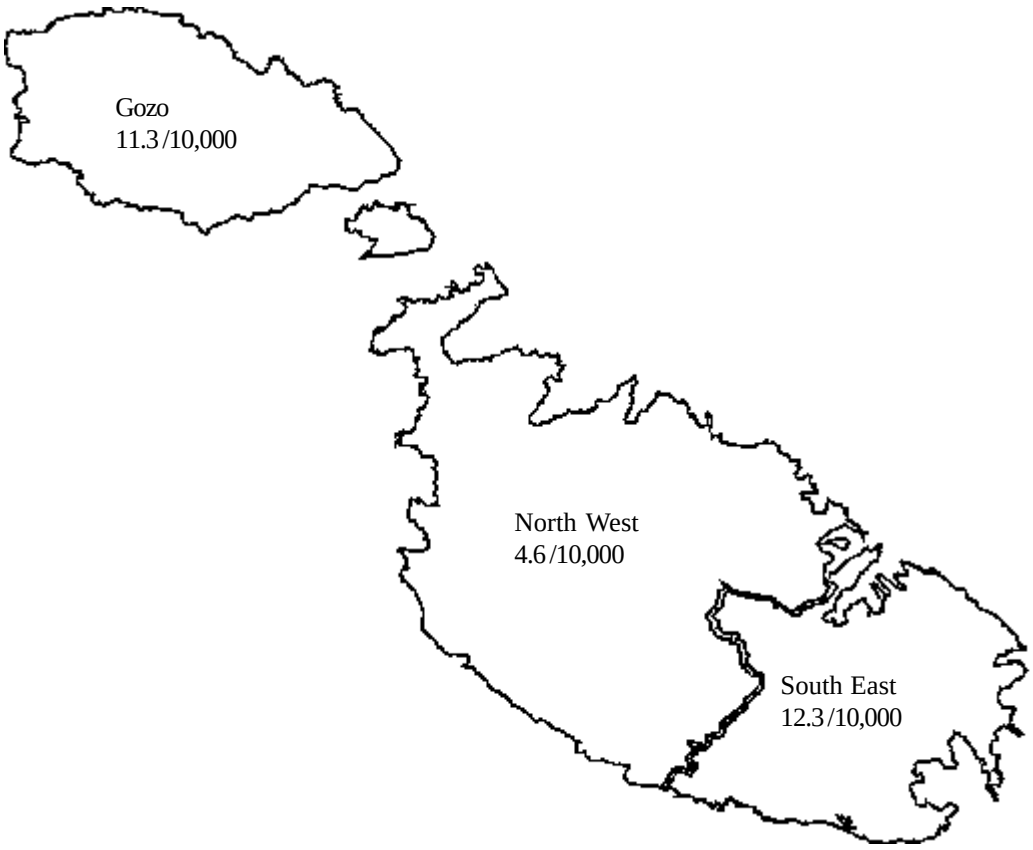
Includes partial or total webbing between two or more digits and includes minor forms (ICD9 code: 7551).

Geographical distribution

Table 41 - Distribution of syndactyly according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**7.5**	**4.0**	**12.2**	**0.0**	**0.0**	**4.6 (1.9 - 10.6)**
Anomalies	2	1	3	0	0	6
Total births	2656	2531	2467	2694	2662	13010
South East	**14.3**	**10.2**	**11.0**	**5.1**	**21.1**	**12.3 (6.7 - 22.2)**
Anomalies	3	2	2	1	4	12
Total births	2102	1955	1822	1957	1896	9732
Gozo	**0.0**	**0.0**	**0.0**	**61.2**	**0.0**	**11.3 (1.2 - 45.5)**
Anomalies	0	0	0	2	0	2
Total births	414	377	344	327	306	1768
Malta & Gozo	**9.7**	**6.2**	**10.8**	**6.0**	**8.2**	**8.2 (5.1 - 12.9)**

Figure 32 - Geographical distribution of syndactyly (1993-97)



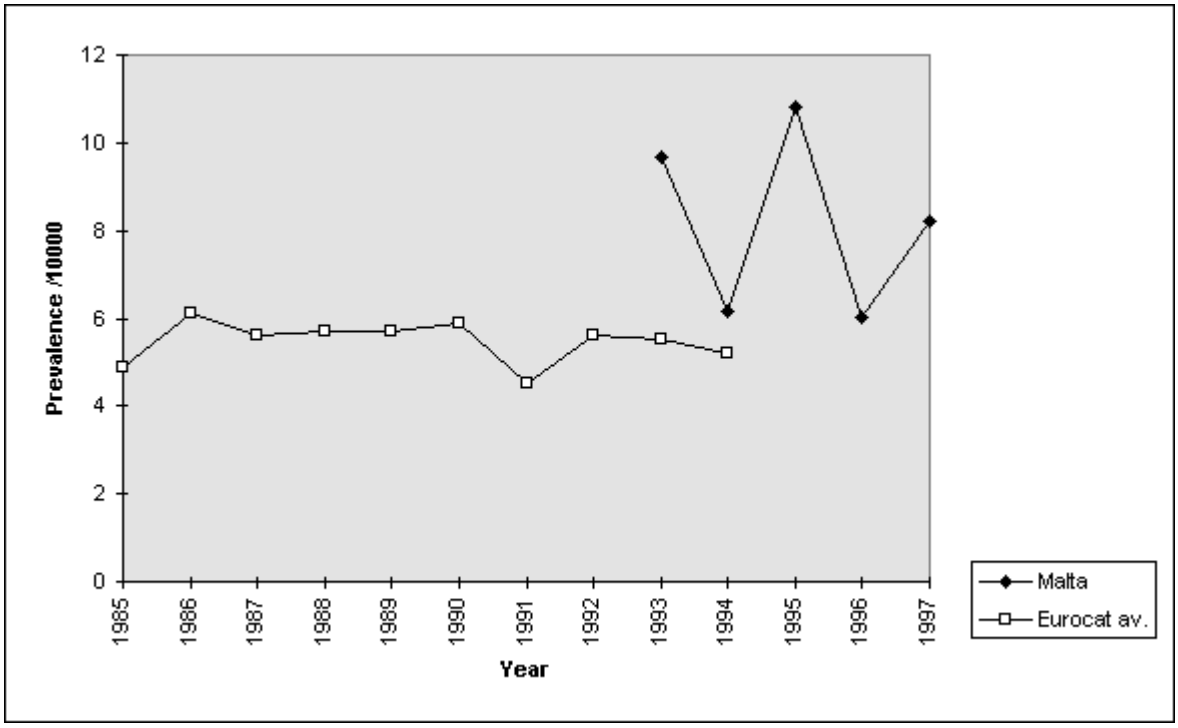
There is a significant difference in prevalence between North West and South East Malta and Gozo with $p<0.05$.

Time trends for syndactyly: Malta & EUROCAT

Table 42 - Time trend of total prevalence of syndactyly: Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	9.7	6.2	10.8	6.0	8.2	8.2 (1993-97)
EUROCAT average*	5.5	5.2				5.8 (1980-94)

Figure 33- Prevalence of syndactyly over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 8.2/10,000 total births (95% CI 5.1 to 12.9)
EUROCAT average 1990-1994 - 5.3/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 0.8* to 8.5** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Galway
** The highest prevalence recorded was in the EUROCAT registry of - Glasgow

DIAPHRAGMATIC HERNIA

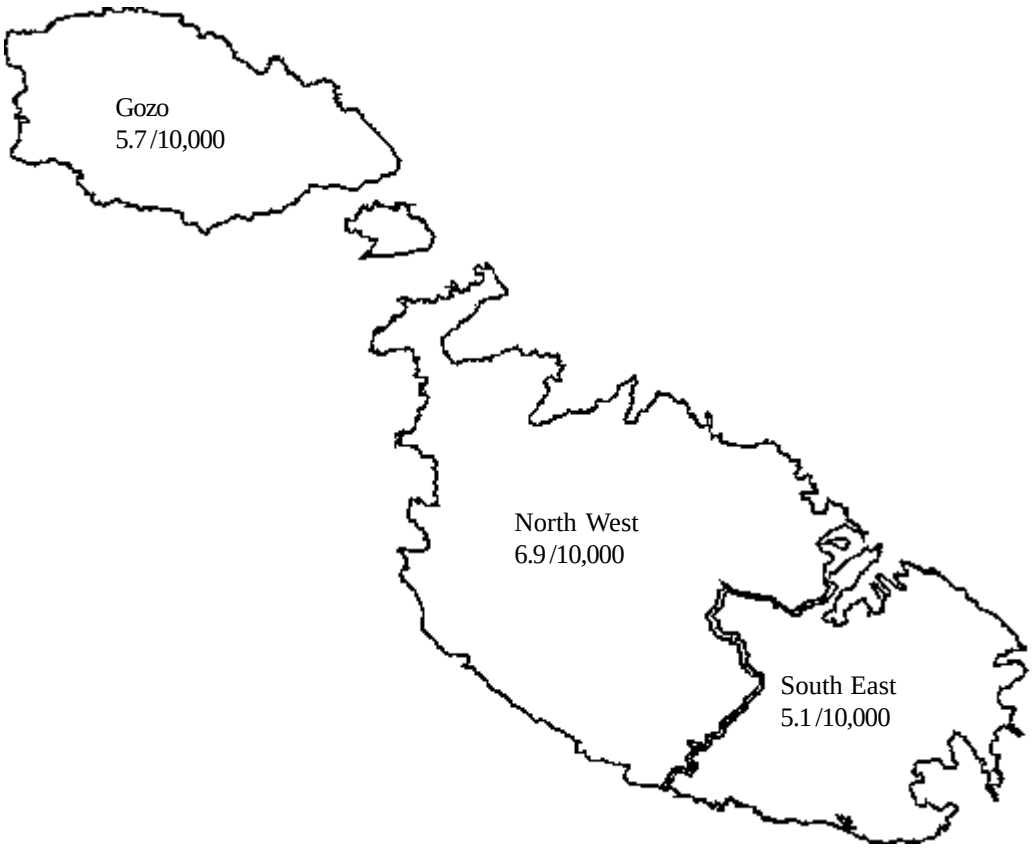
Includes the absence of diaphragm, diaphragmatic hernia, eventration of diaphragm and unspecified anomalies (ICD9 code: 7566)[1].

Geographical distribution

Table 43 - Distribution of diaphragmatic hernia according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**0.0**	**7.9**	**8.1**	**11.1**	**7.5**	**6.9 (3.4 - 13.7)**
Anomalies	0	2	2	3	2	9
Total births	2656	2531	2467	2694	2662	13010
South East	**4.8**	**5.1**	**5.5**	**5.1**	**5.3**	**5.1 (1.9 - 12.7)**
Anomalies	1	1	1	1	1	5
Total births	2102	1955	1822	1957	1896	9732
Gozo	**24.2**	**0.0**	**0.0**	**0.0**	**0.0**	**5.7 (0.3 - 36.7)**
Anomalies	1	0	0	0	0	1
Total births	414	377	344	327	306	1768
Malta & Gozo	**3.9**	**6.2**	**6.5**	**8.0**	**6.2**	**6.1 (3.6 - 10.4)**

Figure 34 - Geographical distribution of diaphragmatic hernia (1993-97)



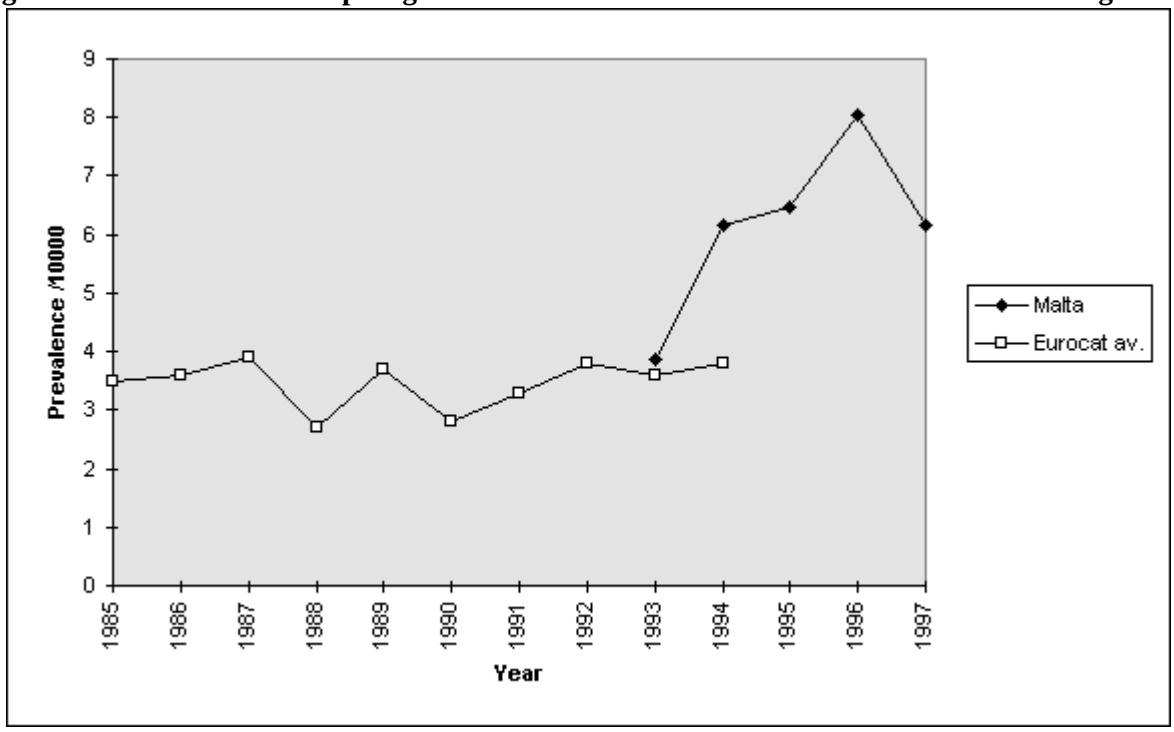
The differences in prevalence are not statistically significant.

Time trends for diaphragmatic hernia: Malta & EUROCAT

Table 44 - Time trend of total prevalence of diaphragmatic hernia: Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	3.9	6.2	6.5	8.0	6.2	6.1 (1993-97)
EUROCAT average*	3.6	6.8				3.4 (1980-94)

Figure 35- Prevalence of diaphragmatic hernia over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 6.1/10,000 total births (95% CI 3.6 to 10.4)
EUROCAT average 1990-1994 - 3.5/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 1.6* to 5.8** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Belfast, Tuscany
** The highest prevalence recorded was in the EUROCAT registry of - Strasbourg

DOWN SYNDROME ALL AGES

Down syndrome / Trisomy 21 is the most frequently encountered chromosomal defect and a major cause of intellectual impairment. Due to the relationship between this defect and maternal age, prevalence is described separately for all maternal ages, maternal ages under 30 years and 35 years or more (ICD9 code: 7580).

Table 45- Distribution of maternal ages for babies with Down syndrome 1993-1997

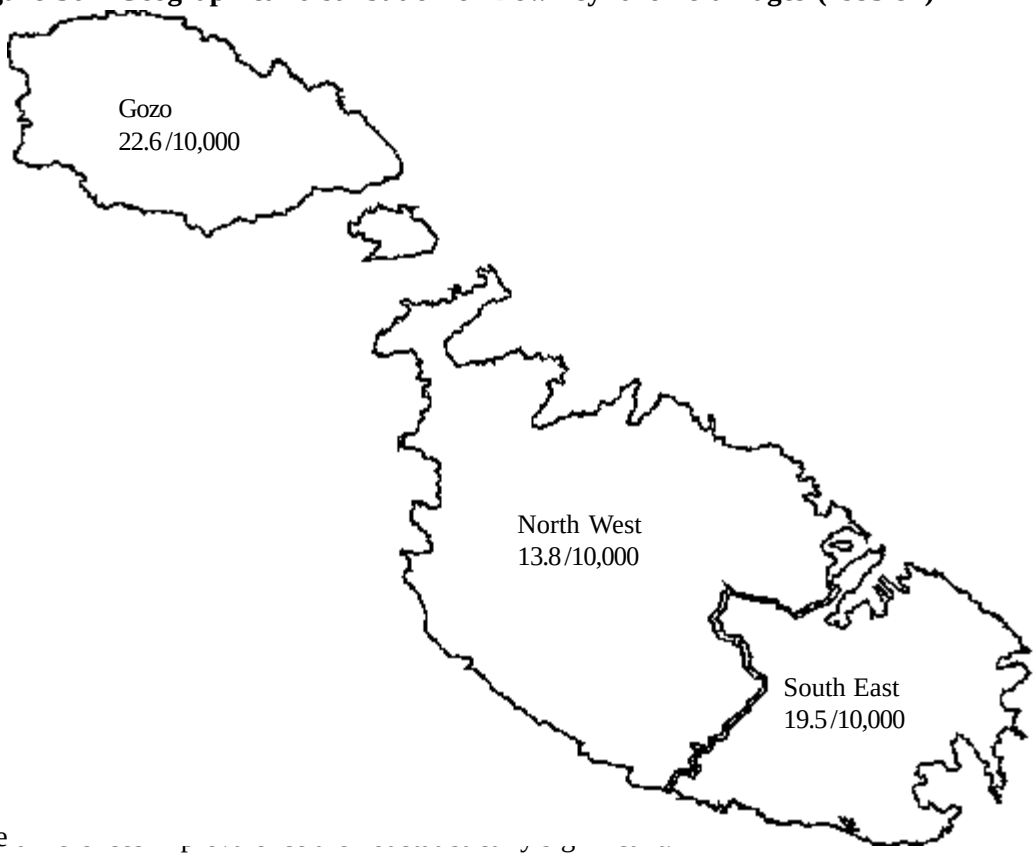
	Maternal age groups							
	<20	20-24	25-29	30-34	35-39	40-44	45+	Total
Babies with Down syndrome	0	0	3	12	17	9	0	41
Total births	957	4932	8581	6489	2910	602	39	24510
Prevalence /10,000	0.0	0.0	3.5	18.5	58.4	149.5	0.0	16.7

Geographical distribution

Table 46 - Distribution of Down syndrome according to locality of residence of mother

Locality	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
North West	**22.6**	**11.9**	**16.2**	**11.1**	**7.5**	**13.8 (8.5 - 22.3)**
Anomalies	6	3	4	3	2	18
Total births	2656	2531	2467	2694	2662	13010
South East	**33.3**	**15.3**	**21.9**	**20.4**	**5.3**	**19.5 (12.1 - 31.1)**
Anomalies	7	3	4	4	1	19
Total births	2102	1955	1822	1957	1896	9732
Gozo	**48.3**	**26.5**	**0.0**	**30.6**	**0.0**	**22.6 (7.3 - 62.1)**
Anomalies	2	1	0	1	0	4
Total births	414	377	344	327	306	1768
Malta & Gozo	**29.0**	**14.4**	**17.3**	**16.1**	**6.2**	**16.7 (12.2 - 22.9)**

Figure 36 - Geographical distribution of Down syndrome all ages (1993-97)



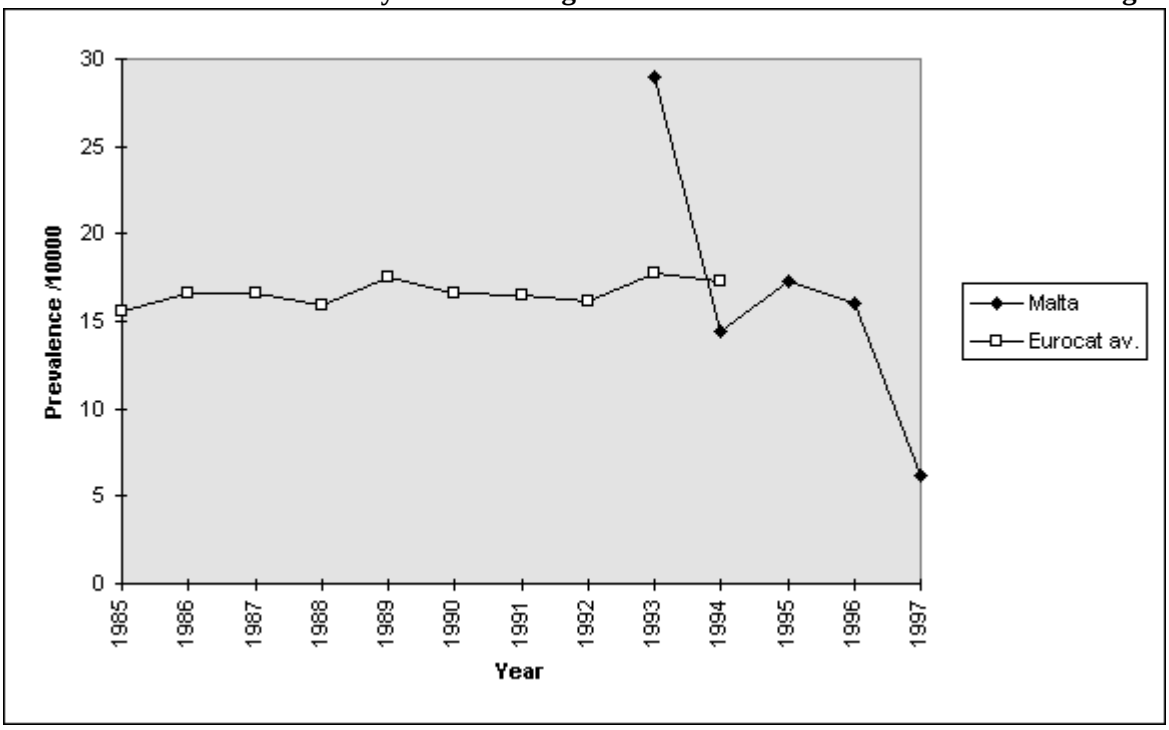
The

Time trends for Down syndrome all ages: Malta & EUROCAT

Table 47 - Time trend of total prevalence of Down syndrome all ages Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	29.0	14.4	17.3	16.1	6.2	16.7 (1993-97)
EUROCAT average*	17.8	17.3				16.1 (1980-94)

Figure 37- Prevalence of Down syndrome all ages over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 16.7/10,000 total births (95% CI 12.2 to 22.9)
EUROCAT average 1990-1994 - 16.8/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 11.5* to 28.0** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Antwerp
** The highest prevalence recorded was in the EUROCAT registry of - Paris

DOWN SYNDROME WITH MATERNAL AGE UNDER 30 YEARS

Time trends for Down syndrome with maternal age under 30 years: Malta & EUROCAT

Table 48 - Prevalence of Down syndrome in Malta with maternal age under 30 years

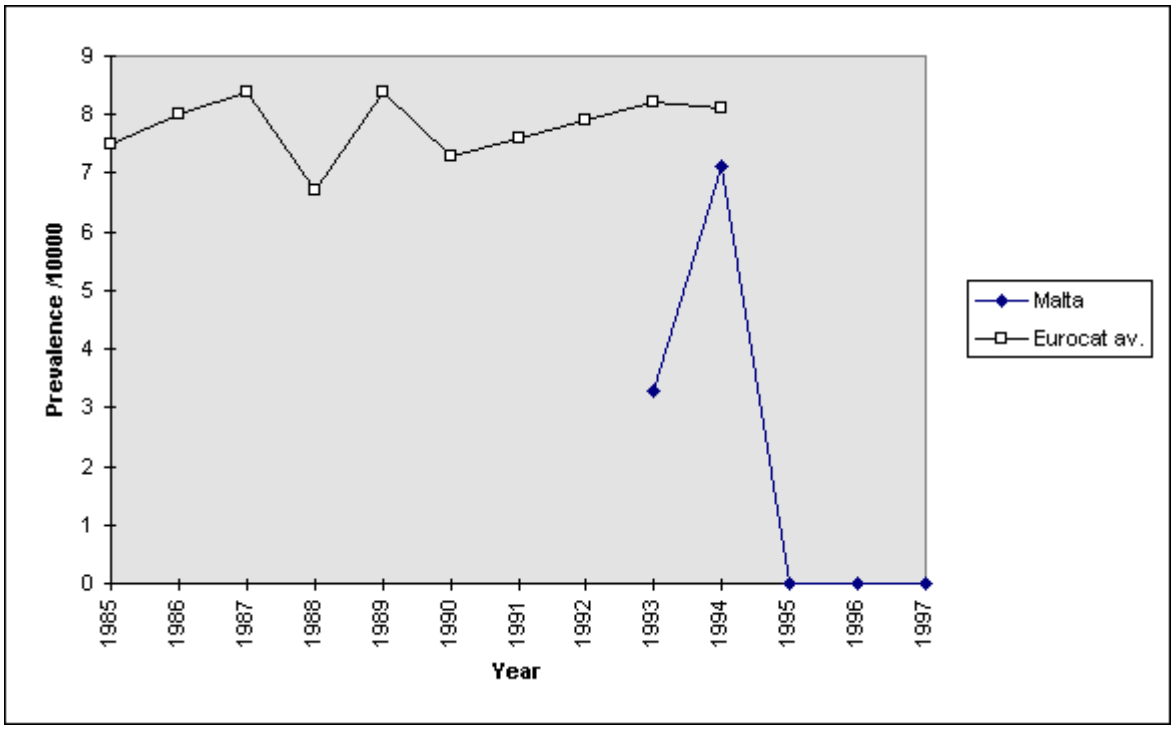
	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
Anomalies	1	2	0	0	0	3
Total births(<30 yrs)	3030	2806	2658	2981	2995	14470
Malta & Gozo	**3.3**	**7.1**	**0.0**	**0.0**	**0.0**	**2.1 (0.5 - 6.6)**

Table 49 - Time trend of prevalence of Down syndrome with maternal age under 30 years Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	3.3	7.1	0.0	0.0	0.0	2.1 (1993-97)
EUROCAT average*	8.2	8.1				7.6 (1980-94)

*Data taken from latest published data: EUROCAT Report 7 [1]

Figure 38- Prevalence of Down syndrome with maternal age under 30 years over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 2.1/10,000 total births (95% CI 0.5 to 6.6)
EUROCAT average 1990-1994 - 7.8/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 5.2* to 11.2** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Odense
** The highest prevalence recorded was in the EUROCAT registries of - Paris, Glasgow

DOWN SYNDROME WITH MATERNAL AGE 35 YEARS OR OVER

Time trends for Down syndrome with maternal ages 35 years and over: Malta & EUROCAT

Table 50 - Prevalence of Down syndrome in Malta with maternal age 35 years and over

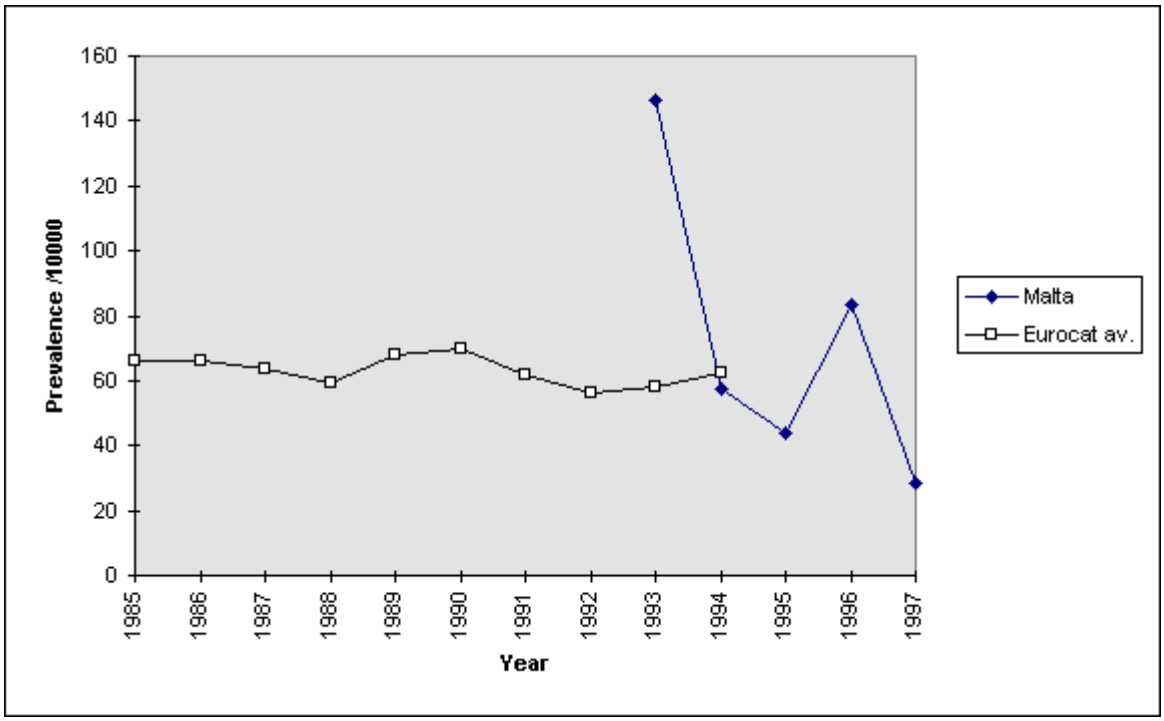
	PREVALENCE /10,000 BIRTHS					
	1993	1994	1995	1996	1997	1993-97 (95% CI)
Anomalies	11	4	3	6	2	26
Total births (>=35 yrs)	751	697	680	718	705	3551
Malta & Gozo	**146.5**	**57.4**	**44.1**	**83.6**	**28.4**	**73.2 (48.9 - 108.7)**

Table 51 - Time trend of prevalence of Down syndrome with maternal ages 35 years and over Malta and EUROCAT average compared

Prevalence/10,000	1993	1994	1995	1996	1997	Total
Malta	146.0	57.4	44.1	83.6	28.4	73.2 (1993-97)
EUROCAT average*	57.8	62.2				61.8 (1980-94)

*Data taken from latest published data: EUROCAT Report 7 [1]

Figure 39- Prevalence of Down syndrome with maternal ages 35 years and more over time Malta and EUROCAT Average



Prevalence in Malta vs EUROCAT

Prevalence in Malta 1993-1997 - 73.2/10,000 total births (95% CI 48.9 to 108.7)
EUROCAT average 1990-1994 - 61.2/10,000 total births

Range of prevalence in EUROCAT registries 1990-1994 - 35.7* to 81.7** /10,000 births
* The lowest prevalence recorded was in the EUROCAT registry of - Antwerp
** The highest prevalence recorded was in the EUROCAT registry of - Paris

REFERENCES

[1] A EUROCAT Working Group. 15 years of Surveillance of Congenital Anomalies in Europe 1980-1994 - Report 7 and Addendum. Brussels, Institute of Hygiene and Epidemiology, 1997: pp 10-149.

[2] Demographic Review for the Maltese Islands for the years 1993 to 1997. Malta: Central Office of Statistics (Annual Publications).

[3] British Paediatric Association Classification of Diseases. London, The British Paediatric Association, 1979: pp 1-220.

[4] EUROCAT Central Registry. EUROCAT Guide 1: Instructions for the Registration of Congenital Anomalies. Brussels, Department of Epidemiology, Catholic University of Louvain, 1990: pp 1-59.

ANNEX 1 LOCAL REPORT FORM

CONFIDENTIAL REPORT ON CONGENITAL ANOMALIES

CHILD'S NAME: _____ I.D._____

ADDRESS: _____
_____
_____

DATE OF BIRTH: _____

MOTHER'S NAME: _____ I.D._____

DIAGNOSIS: _____
_____
_____
_____
_____

REFERRING DOCTOR:_____ DATE:_____

PLEASESEND INFORMATION TO DR. MIRIAM GATT, DEPT. OF HEALTH INFORMATION, 95 G'MANGIA HILL, G'MANGIA MSD 08

ANNEX 2 LIST OF EXCLUSIONS

LIST OF MINOR ANOMALIES (AND ICD-9 CODES) WHICH ARE NOT INCLUDED IN REPORTS UNLESS IN COMBINATION WITH OTHER MAJOR ANOMALIES[1]

Anomalies of the eye
- Stenosis or stricture of lacrimal duct (74365)

Anomalies of the ear
- Minor or unspecified anomaly of the ear (7443)
- Preauricular appendage, tag or lobule (74411)
- Other appendage tag or lobule (74412)

Cardiovascular system
- Functional or unspecified cardiac murmur (7852)
- Absence or hypoplasia of umbilical artery, single umbilical artery (7475)
- Patent ductus arteriosus (in babies < 37 weeks or < 2500g) (7470)

Digestive system
- Tongue tie (7500)

External genitalis
- Undescended testicle (7525) and unspecified ectopic testis (75253)
- Congenital hydrocele or hydrocele of testis (7786)
- Phymosis (605)
- Hypospadias when the meatus lies before the coronary sulcus, glandular or 1st degree hypospadias (75260)

Limbs
- Clicking hip (75432)
- Clubfoot of postural origin (75473)
- Postural or unspecified metatarsus varus or metatarsus adductus (75452)
- Postural or unspecified talipes calcaneovalgus or pes calcaneovalgus (75460)
- Minor or unspecified anomalies of toe such as hallux valgus, hallux varus or “orteil en marteau” (75560)

Other musculoskeletal anomalies and anomalies of the integument
- Spina bifida occulta uncomplicated (75610)
- Pectus excavatum (75636 or 75481)
- Minor or unspecified anomaly of the nose (74819)
- Minor or unspecified deformity of the face (74491)
- Minor anomaly of nipple (75768)
- Accessory or ectopic nipple (75765)
- Congenital umbilical hernia (5531), inguinal hernia (550), para umbilical (5531), ventral or incisional (5532), hiatus hernia (7506)
- Abnormal palmar crease (7572)
- Skin tag with surface area < 4 cm^2 : skin tag (75731), naevus (75738), angioma (2280), haemangioma (2280), glomus tumor (2280), lymphangioma (2281), birthmark (75738)
- Sacral dimple (7578 or 6851)

ANNEX 3 EUROCAT SUBGROUPS

EUROCAT subgroup and Text		ICD-9 codes included
01	Neural Tube defects	7400-7420
02	Congenital Hydrocephaly	7423 excl 74232
03	Microcephaly	7421,
04	Other Congenital anomalies of the Nervous System	7422, 7424-5, 7428-9, 3440-9, 3492, 3519, 3526, 225-, 2375-7, 2370-1, 2379
05	Congenital anomalies of the eye	7430-7439, 3627, 3622, 3743, 3787, 3795, 224
06	Congenital anomalies of the ear	7440-7443
07	Anomalies of cardiac septal closure	7450-7459
08	Other congenital anomalies of the heart	7460-7469, 4253, 7852,
09	Other congenital anomalies of circulatory system	7470-6, 7478, 7479
10	Anomalies of the Respiratory System	7482-6, 7488-9, 2358, 7702, 4920
11	Cleft lip and palate	7490-7492
12	Tracheo-oesophageal fistula, oesophageal atresia and stenosis	75030-75038,
13	Atresia and stenosis of large intestine, rectum and anal canal	75121-75124
14	Other anomalies of the digestive system and upper alimentary tract	7504-5, 7507-11, 7413-9, 75120, 211-
15	Anomalies of the external genital organs (male)	7525-6, 7528, 7786
16	Anomalies of external female genitalia and indeterminate sex	7524, 7527
17	Other anomalies of genital organs	7520-3, 7529, 219-, 220-, 221-, 222-,
18	Renal Anomalies	7530-1, 7533, 2330,
19	Anomalies of urinary tract	7532, 7534-9, 2231-3, 2238-9
20	Polydactyly and syndactyly	7550-7551
21	Reduction deformities of limbs	7552-7554
22	Talipes	7545-7547
23	Other anomalies of limbs	7543-4, 7555-6, 7558-9
24	Anomalies of the tongue, brachial cleft, auricular sinus	7444, 7500-2
25	Anomalies of nose, face, skull and neck	5240, 5249, 7445, 7448-9, 7480-1, 7540, 7560, 5209
26	Anomalies of the diaphragm	7506, 7566
27	Other musculoskeletal anomalies of thorax and neck	7548, 7561, 7541-2, 7562-3
28	Anomalies of the abdominal wall	7567, 5509, 5530-2, 5538-9
29	Other anomalies of bones, muscles, cartilages and connective tissues	7564-5, 7568-9, 2380
30	Anomalies of the integument	7570-2, 7574-5, 7578-9
31	Skin tags and benign neoplasms of the skin	6851, 7573, 2140, 2169, 2280-1
32	Down Syndrome	7580,
33	Other chromosomal anomalies, Down Syndrome excluded	7581-9
34	Metabolic defects	2439, 2461, 2533-4, 2552, 2579, 2598, 2700-13 2718-33, 2738-41, 2748-54, 2758-59, 2770-6, 2778-79, 2820-7, 2860-3
35	Anomalies of breast, spleen, adrenal gland and other endocrine glands	7576, 7590-2
36	Other syndromes, eponyms and disease names	2873, 2879, 7593-6, 7598, 7602, 7710-2, 0900, 8888, 7726
37	Other unspecified and ill-defined conditions	7599, 7597, 7780
38	Other	Conditions NEC

Centre No. [2][3]

EUROCAT REPORT ON CONGENITAL ANOMALIES

INFANT
Local ID No.

Date of Birth
Place of Birth
Sex (1 = male, 2 = female, 3 = indeterminate, 9 = nk)
No. of babies delivered
Birth order (in multiple set)
No. malformed (in multiple set)
Type of Birth (1 = live, 2 = still, 3 = spont. abortion, 4 = induced abortion, 9 = nk)
Birth weight (grams)
Date of L.M.P. and Certainty
Length of gestation (weeks)
Date of death
Survival beyond a week of age (1 = yes, 2 = no, 9 = nk)
Sources of information

DIAGNOSIS - TIME & TECHNIQUES
Date of discovery
When discovered*
If prenatally diagnosed, gest. age at discovery
Condition at discovery (1 = live, 2 = dead, 9 = nk)
Prenatal diagnosis (1 = done, result positive, 2 = done, result nk, 3 = not done, 4 = result negative, 8 = N/Ant, 9 = nk)
- Amniocentesis
- Ultrasound
- Chorionic Villi sampling
- Other techniques (specify) _____

Karyotype of infant/fetus (specify) (1 = done not k, 2 = done ca, nk, 3 = not done, 8 = N/Ant, 9 = nk) .....
Post mortem examination (1 = done not k, 2 = done nk, 3 = not done, 4 = autopsy done, 9 = nk)

MOTHER
Residence code
Date of birth
Age at delivery (years)
Reproductive history
- Prev. spontaneous abortion n°
- Prev. induced abortion n°
- Prev. live birth(s) n°
- Prev. stillbirth(s) n°

Total Prev. pregnancies n°
Occupation _____
Social status
Racial type
Assisted conception * _____
Illness before pregnancy * _____
Illness during pregnancy * _____
_____
Habitual exposures * _____
(specify) _____
Unusual exposure (specify) _____
Drugs (1st trim) 1. _____
2. _____
3. _____

FATHER
Date of birth
Age at delivery (years)
Occupation _____
Social status
Racial type
Chronic illness

DIAGNOSIS - MALFORMATIONS
Syndrome *
Malformations present
1. _____ *
2. _____ *
3. _____ *
4. _____ *
5. _____ *
6. _____ *
7. _____ *
8. _____ *

McKusick Code/Type of Mendelian inheritance
Mode of transmission (for single gene or chromosomal disorders, 1 = familial, 2 = de novo, 9 = nk)
Consanguinity *
Prev. sibs notified to EUROCAT (1 = yes, 2 = no, 9 = nk)
Local code No. 1.
2.
3.
Family history of anomaly
Sibs with anomaly * (Specify) _____
Mother's family * (Specify) _____
Father's family * (Specify) _____
Systems involved _____
Category _____

__/__/__

* : see instructions *Italics : for local use only*

Enter additional malformations / comments below:

Mother interviewed _____
Mother/Baby files seen _____

DH 1018

Name _____
Address _____
_____

ANNEX 5 GEOGRAPHICAL REGIONS

GEOGRAPHICAL REGIONS ACCORDING TO DEPARTMENT OF HEALTH INFORMATION DIVISIONS

North-West group of regions				
West	North	Central	Central North	Central West
Zebbug	Mellieha	Hamrun	Sliema	Attard
Siggiewi	Mgarr	Sta Venera	St. Julian's	Balzan
Mdina	St Paul	Msida/G'Mangia/Pieta	San Gwann	Birkirkara
Dingli	Gharghur	Gzira/Ta' Xbiex		Lija/Iklin
Rabat	Mosta			
	Naxxar			

South-East group of regions				
Central East	Harbour	Central South	South	East
Fgura	Valletta	Qormi	Gudja	Zejtun
Paola	Floriana	Marsa	Kirkop	Birzebbugia
Sta Lucia	Vittoriosa		Luqa	Ghaxaq
Tarxien	Senglea		Mqabba	Marsaxlokk
	Cospicua		Qrendi	Zabbar/Xghajra
	Kalkara		Safi	Marsascala
			Zurrieq	