EUROCAT GUIDE-LINES

FOR

REGISTRATION OF CONGENITAL ABNORMALITIES

AND

MULTIPLE BIRTHS

Edited by J.A.C. Weatherall

Project Leader

1980

An EEC Concerted Action Project

GUIDE - LINES

to the

REGISTRATION OF CONGENITAL ABNORMALITIES

AND

MULTIPLE BIRTHS

FOR

EUROCAT

an

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I. INTRODUCTION

In 1974 the Committee of Medical and Public Health Research (CRM) of the EEC decided to promote medical research by Concerted Action. Research activities within each country concerning several particular topics would, for each topic, be coordinated to a central plan. Cost of research within each country would be financed by the Member States ; cost of coordination of the research would be provided and financed by the Commission. Working groups of the CRM were invited to prepare projects for Concerted Action and the Working Group in Epidemiology, Statistics and Clinical Trials put forward a project in the field of epidemiology which consisted of establishing registers, of congenital malformations and multiple births.

After a feasibility study conducted in 1975 and 1976 the Concerted Action on Congenital Abnormalities and Multiple Births was established by the decision of 13th February 1978 (Ref.OJ. n° L52, 23.2.1978, page 20). The study is supervised for the commission by a steering committee (COMAC) whose members are nominated by the participating states (current members are shown in Appendix I).

Initially 15 study areas were proposed within 9 countries. In 1979, the participation of Greece with an additional area was officially approved. Other centres within the EEC countries and outside the EEC will possibly join the study. Each local register area is directed by a registry leader (Appendix II) who is responsible for day to day running of each study.

The coordination financed by the Commission is carried out by a project leader who performs this task from a base in the Department of Epidemiology at the School of Public Health in the University of Louvain in Brussels (Ecole de Santé Publique, B.P. EPID 30.34, Clos Chapelle-aux-Champs 30, 1200 Bruxelles - Belgium).

The Concerted Action Project in Registration of Congenital Abnormalities and Twins has been named EUROCAT.

II. OBJECTIVES OF THE STUDY

The long term objective of this study is to test the feasibility of carrying out epidemiological surveillance in the countries of the E.E.C. taking surveillance of congenital abnormalities and multiple births as an example.

The specific objectives of the study are :

1. To set up within each selected area in each country a population based register of congenital abnormalities and multiple deliveries. In order to achieve full recording, ideally the outcome of every conception to women resident in the defined area should be known. This involves searching for congenital malformations, biochemical and chromosomal anomalies in aborted fetuses, in live and stillborn infants, in dead children and during childhood. Babies from multiple deliveries should be recorded at birth.

2. To study the methods of data collection in each centre, to evaluate the effect of these in biasing the data collected and to propose and test ways to circumvent these difficulties.

3. To monitor the incidence rates reported in different population groups at different times in order to identify possible etiological factors.

4. To create in each country an area where reporting is reliable, so that base line rates are available for use in calibrating any national warning system established for the detection of adverse environmental influences by allowing the interpretation of a reported increasing rate.

5. To evaluate the effectiveness and efficiency of screening programs and preventive measures.

6. To provide a well documented set of individuals recorded in a defined population for further specific studies, such as follow up studies of cases with specified malformations in order to compare the results of different treatment regimens.

7. To establish the means by which multiple births can be efficiently registered at birth and how information can be collected which will allow the reliable and cheap recording of zygosity.

III. CASES TO BE RECORDED AT LOCAL LEVEL

All pregnancies ending in either abortion or birth to mothers resident within the selected area, which take place inside or outside the selected area, should be included in the survey and will form the denominator of incidence rates.

The following cases will be registered : any baby or fetus ;

- born of multiple pregnancy ;

or which have either :

- one or more congenital malformations ;
- any chromosomal anomaly ;
- any metabolic anomaly affecting the child from birth onwards.

As there is no general agreement about the definition of congenital malformation, local physicians should be asked to notify to their register anything they consider to be a malformation. This is recommended with the intention of making the detection as sensitive as possible even if trivial conditions are over-reported. It is easier to exclude notified cases with trivial conditions than to search for unreported cases of severe malformations.

There is no limit to age at registration. Cases can be discovered:

1. When a pregnancy ends in a spontaneous or legal abortion ;
2. At birth of a live or stillborn child ;
3. During the early neonatal period (from birth to 7 days) ;
4. At any age after the first week of life.

It is expected that all registers will, in the first instance, achieve a complete coverage of cases discovered at birth or during the early neonatal period. For all cases it is important to indicate the age of the child at the time he was recognized as malformed.

In the first instance, follow up of all malformed children up to one year should be established in order to exclude falsely diagnosed cases. Follow up through childhood should also be considered so that twins and children surviving with malformations can be available for studies if they are necessary.

IV. INFORMATION TO BE RECORDED ABOUT EACH CASE, AT LOCAL LEVEL

Each local register will record the identity and address of the cases. At local level, the use of named records is needed (1) to link reports arriving from several sources and so, to avoid a multiple registration ; (2) to allow the follow up of cases which is required to ascertain the diagnosis and to study the outcome of malformed children ; (3) to trace the cases in order to conduct prospective or retrospective studies.

A limited list of data about the case, its mother and father (Appendix III), is to be transmitted to EUROCAT, along with the diagnosis of all malformations and a local number for each case.

It is understood that some centres have a special interest in particular topics and are already recording other specific information which will continue to be collected in addition to the information needed for EUROCAT.

V. CASES AND INFORMATION TO BE TRANSMITTED TO EUROCAT

All the cases recorded at local level with single or multiple malformations included in chapter 14 of the International Classification of Diseases, 9th Edition, vol. 1., or are named "congenital", "inherited", "inborn error" in other chapters, are to be transmitted to EUROCAT.

Since a lot of minor and isolated malformations are considered to be redundant, it is recommended not to transmit those cases that are included in the annexed list (Appendix IV).

The cases, for which the diagnosis of malformation is well established, are to be transmitted MONTHLY to EUROCAT. The cases which are certainly malformed but for which the exact diagnosis is not yet certain should also be transmitted to EUROCAT. When further information is available about a case already notified an amending report is to be transmitted. The cases, for which a malformation is only suspected, are to be recorded locally and transmitted to EUROCAT only when the diagnosis is confirmed. If after a year, the suspicion has not been confirmed or the case has been lost to follow up, the cases should be transmitted to Central Registry as a separate batch.

The names and addresses of cases and hospitals are to be kept at the local registry department and will not be sent to EUROCAT. The local identity number, that is unique for each baby, is to be transmitted to EUROCAT. In order to achieve confidentiality and data security, this local identity number will not be added to the central computer record for the child. EUROCAT will allocate EUROCAT's own serial number to each case. This will be used in the computer. The linkage will be done manually.

A standard pre-coded transmission form (Appendix V) and the appropriate coding instructions (Appendix VI) are proposed.

If, due to local convenience, this standard form cannot be adopted, any other transmission form may be used, providing the data are compatible.

For the written information in the completed form, the use of French or English is requested.

APPENDIX I : LIST OF COMAC MEMBERS

NAME	COUNTRY	ADDRESS
Prof. M.F. LECHAT (Chairman)	Belgium	Université Catholique de Louvain Ecole de Santé Publique EPID. 30.34 Clos Chapelle-aux-Champs, 30 B - 1200 Bruxelles
Prof. M. HAUGE	Denmark	Arvepatologish Institut Odense Universitet J.B. Winslowsvej 17 DK - Odense C
Dr C. RUMEAU-ROUQUETTE	France	Groupe de Recherches d'Epidémiologie sur la Mère et l'Enfant 44, Chemin de Ronde F - 78110 Le Vésinet
Dr J. FEINGOLD	France	Groupe de Recherches de Génétique Epidémiologiques Château de Longchamp F - 75016 Paris
Dr K.H. DEGENHARDT	Germany	Institut für Humangenetik der Universität Frankfurt 41, Paul Ehrlischstrasse D - 6000 Frankfurt/Main
Dr S. TSAGARAKI	Greece	Children's Hosp. Agia Sophia Institute of Child Health Athens 617
Prof. TENTORI	Italy	Istituto Superior De Sanita Viale Regina Elena 299 I - 00161 Roma
Dr G. DEAN	Ireland	The Medico-Social Research Board 75, Lower Baggot Street IRL - Dublin
Dr T. O'DWYER	Ireland	Department of Health Custom House IRL - Dublin I
Dr A. BETZ	Luxembourg	Institut d'Hygiène et de la Santé Publique Rue de Laboratoire, 42 L - Luxembourg

Dr W.M.J. VAN DUYNE	The Netherlands	Nerderlands Instituut voor Preventive Geneeskunde Postbus 124 NL - Leiden
Dr G.FORWELL	United Kingdom	Greater Glasgow Health Board 321 Sauchiehall Street GB - Glasgow G 2 3HT
Dr R.W.SMITHELLS	United Kingdom	Depart. of Paediatrics and Child Health - University of Leeds 27, Blundell Street GB - Leeds LS1 3ET

APPENDIX II : LIST OF PARTICIPATING REGISTERS AND REGISTRY LEADERS

NAME OF REGISTER	COUNTRY	REGISTRY LEADER	ADDRESS
Hainaut	Belgium	Dr I. BORLEE	Ecole de Santé Publique UCL - EPID 30.34 Clos Chapelle-aux-Champs 30 B - 1200 Bruxelles Tél : (02) 762.34.00 Ext: 3325
W. Vlaanderen	Belgium	Prof R. BECKERS	Ministerie van Volksgezonheid en van het Gezin Administratief Centrum Vesaliusgebouw B - 1010 Brussel Tél : (02) 564.80.11 prive: (050) 33.21.82
Odense	Denmark	Dr M. ULRICH	University of Odense Dept. of Clinical Genetics 17, J.B. Winslowsvej DK - 5000 Odense C Tél : (09) 12.71.79
Frankfurt	Germany	Prof K.H. DEGENHARDT	Institut für Humangenetik der Universitat 41 Paul Ehrlischstrasse D - 6000 Frankfurt#Main Tél : Frankfurt 63.01.60
Evia	Greece	Dr S. TSAGARAKI	Children's Hospital Agia Sophia Institute of Child Health GR - Athens 617 Tél : Athens 7708291
Firenze	Italy	Dr C. GALANTI	Assessorato Della Sanita Dipartimento Sicurezza Sociale 26 Via di Novoli I - FIRENZE CAP 50100 Tél : (055) 27.661
Roma	Italy	Dr P. MASTROIACOVO	Istituto di Pediatria University Cattolica Largo Agostino Gemelli 8 I - 00168 ROMA Tél: Rome 33051

Morlaix	France	Dr GIRARD	Département de Pédiatrie Hôpital de Morlaix F - Morlaix (Finistère)
Paris-Yvelines	France	Dr (Mme) J. GOUJARD	Groupe de Recherches Epidémiologique sur la Mère et l'Enfant I.N.S.E.R.M. 44 Chemin de Ronde F - 78110 Le Vésinet Tel : Paris 976 3333
Dublin	Ireland	Dr RADIC	The Medico-Social Research Board 73 Lower Baggot Street IRL - DUBLIN 2 Tél : (01) 76.11.76 (01) 76.60.76
Galway	Ireland	Dr P. POWER	Director of Community Care and Medical Officer of Health Hiberian Building, Eyre Square IRL, GALWAY
Luxembourg	Luxembourg	Dr D. HANSEN-KOENIG	Ministère de la Santé Publique Division de Médecine Préventive et Sociale 10 av. de la Liberté L - LUXEMBOURG Tél : 40801
Groningen	The Netherlands	Dr L.P. TEN KATE	Dept. of Human Genetics University of Groningen 4, A. Deusinglaan NL - Groningen
Glasgow	United Kingdom	Dr F. HAMILTON	Greater Glasgow Health Board 351 Sauchiehall Street GB - Glasgow G2 3HT Tél : (041) 332.29.77
Liverpool	United Kingdom	Prof F.HARRIS	Institute of Child Health Dept. of Child Health Alder Hey Children's Hospital Eaton Road GB - Liverpool L12 2AP Tél : (051) 228 2024
Belfast	United Kingdom	Prof N.C.NEVIN	The Queen's University of Belfast Dept. of Medical Genetics Institute of Clinical Science Grosvenor Road GB - Belfast BT12 6BJ Tél : 40.503 ext : 392

APPENDIX III: LAYOUT OF EUROCAT REGISTER ON CONGENITAL ABNORMALITIES AND TWINS

1-47 DATA ABOUT THE CASE

1-11 EUROCAT Serial Number (to be coded centrally)
- 1-2 : Registry Code Number
- 3-9 : Arrival Number
- 10-11 : Check Number

12 Card Number = [1]

13-18 Date of birth (Day-Month-Year) 999999 = N/S
- 2 digit day (01-31)
- 2 digit month (01-12)
- 2 digit year (last 2 digits of year)

19-23 Place of birth (local code)
99999 : N/K

24 Sex 1 = Male 3 = Indeterminate
2 = Female 9 = N/K

25 Numbers of babies delivered
1 = Singleton 3 = Triplet 9 = N/K
2 = Twins

26 Type of Birth
1 = LB 3 = SA 9 = N/K
2 = SB 4 = IA

27-30 Birth weight (in grams)
9999 : N/K

31-36 Date of L.M.P. (Day-Month-Year)
- 2 digit day (01-31)
- 2 digit month (01-12)
- 2 digit year (last 2 digits of year)

000000 : NO L.M.P. 999999 : N/K

37-38 Length of gestation (in completed weeks)
00 : impossible to ascertain 99 : N/S

39 Certainty of LMP 1 = certain
2 = uncertain 9 = N/K or N/S
0 or 3 = NO LMP

40 Prenatal diagnosis
41 Karyotype
42 Post Mortem examination

0 or 3 = not performed
1 = performed, result known
2 = performed, result unknown
9 = N/K or N/S

43-47 Source of Information

1 : Notes in routine scan
2 : Birth notification or notification of malformation at birth
3 : Hospital case notes
4 : Death or Stillbirth certificate
5 : Pre-natal diagnosis report
6 : Laboratory report (cytogenetic, chemical, etc...)
7 : Post mortem examination
8 : Other
9 : N/S

43 1st source of information
44 2nd source of information
45 3rd source of information
46 4th source of information
47 5th source of information

48-53 MULTIPLE BIRTHS (This section is to be completed only for multiple birt

48 Birth order
1,2,3 etc. 9 = multiple birth, order unknown

49 Number of malformed babies/fetuses
1,2,3 etc. 9 = multiple birth and not stated

50 Sex
1 : like sex 3 : indeterminate 9 : multiple, N/S
2 : unlike sex 8 : other

51 Chorionic membrane
1 : monochorial 8 : other
2 : dichorial 9 : if multiple, N/S

52 Amniotic membrane
1 : monoamniotic 8 : other
2 : diamniotic 9 : if multiple, N/S

53 Other
1 : yes 9 : N/K or N/S
2 : no

1-50 DATA ABOUT THE MOTHER

1-11 EUROCAT Serial Number (to be coded centrally)
- 1-2 : Registry Code Number
- 3-9 : Arrival Number
- 10-11 : Check Number

12 Card Number = [2]

13-19 Residence Code (local code)
9999999 : N/S

20-25 Date of Birth (Day-Month-Year)
- 2 digit day (01-31)
- 2 digit month (01-12)
- 2 digit year (last 2 digits of year)

999999 : N/S

26-27 Age at delivery (in completed years)
99 : N/S or N/K

28 Number of spontaneous abortions
9 : N/S or N/K

29 Number of induced abortions
9 : N/S or N/K

30-31 Number of live births
99 : N/S or N/K

32 Number of stillbirths
9 : N/S or N/K

33-34 Total number of previous pregnancies
99 : N/S or N/K

35-39 Occupation (to be coded locally)
99999 = N/S or N/K

40 Social status (to be coded locally)
9 : N/S

41 Racial type (local code)
9 : N/S

42 Previous child(ren) with anomaly(ies)

43 Family history of anomaly (ies)

44 Chronic illness

45 Illness during pregnancy

46 Smoking

47 Alcohol

48 Drugs during pregnancy

49 Family history of twinning

1 : YES
0 or 2 : NO
9 : N/K or N/S

50 Consanguinity

0 : not related
1 : 1st cousin
2 : 2nd cousin
8 : other relation or relation not stated
9 : N/K

51-71 DATA ABOUT THE FATHER

51-56 Date of birth (Day-Month-Year)
2 digit day (01-31)
2 digit month (01-12)
2 digit year (2 last digits of year)
999999 = N/S

57-58 Age at Birth (in completed years)
99 : N/S

59-63 Occupation (to be coded locally)
99999 : N/S

64 Social status (to be coded locally)
9 : N/S

65 Racial type (local code)
9 : N/S

66 Family history of anomaly (ies)
67 Chronic illness
68 Smoking
69 Alcohol
70 Drugs
71 Family history of twinning

1 = YES
0 or 2: NO
9 = N/K or N/S

1-78 MALFORMATIONS

1-11 EUROCAT Serial Number (to be coded centrally)
1-2 : Registry Code Number
3-9 : Arrival Number
10-11 : Check Number

12 Card Number = [3]

13-18 Date of discovery (Day-Month-Year)
2 digit day (01-31)
2 digit month (01-12)
2 digit year (last 2 digits of year)
999999 = N/S

19 When discovered
1 : birth
2 : less than 1 week
3 : 1-4 weeks
4 : 1-12 months
5 : over 12 months
6 : prenatal diagnosis
7 : spontaneous or induced abortion
9 : N/S or N/K

20 Condition at discovery
1 : live
2 : dead
9 : N/K

21-26 Syndrome, eponym or disease name
ICD, BPA or EUROCAT code

27-32 1st malformation
33-38 2nd malformation
39-44 3rd malformation
45-50 4th malformation
51-56 5th malformation
57-62 6th malformation
63-68 7th malformation
69-74 8th malformation
} ICD or BPA code

75 Ascertainment of diagnosis
0 = no malformation
1 = confirmed
2 = still doubtful
3 = lost to follow-up
9 = N/K

76 Mode of inheritance
0 : autosomal dominant
1 : autosomal recessive
2 : X-linked recessive/dominant
3 : numeric chromosomal defect of autosomes
4 : structural chromosomal defect of autosomes
5 : numeric sex chromosomal defects
6 : structural sex chromosomal defects
7 : polygenic/multifactorial
8 : known environmental agent
9 : other or unknown

77-78 Number of malformations : to be coded centrally

APPENDIX IV

CASES FOR EXCLUSION OR SPECIAL CONSIDERATION

Reports of cases with the following minor malformations are not to be transmitted to EUROCAT unless occuring in combination with other specified abnormalities. (Provisional list).

Uncomplicated Spina Bifida Occulta (756-10). This is usually diagnosed by radiography and usually later in childhood (see definition in CNS meeting report).

Stenosis or stricture of lacrimal duct (743.65).

Absence or hypoplasia of umbilical artery (single umbilical artery) (747.5).

Unspecified anomalies (deformities) of ear (744.3).

Unspecified anomalies (deformities) of nose (748.19) without further information.

Undescended testicle (752.5), unilateral
Undescended testicle (752.50), bilateral
Undescended testicle (752.51), undescended testicle, NOS (752.52), and not specified ectopic testis (part of 752.53).

Reductant prepuce and phimosis (605)
Congenital hydrocele or hydrocele of testis (778.6).
Breat engorgement in newborn (778.7).

Small anomalies of toes such as hallux valgus, hallux varus or "orteil en marteau" (755.60).

Single hernia - umbilical (553.1)
- inguinal (550.-)
- para umbilical (553.1)

Abnormal palmar creases (757.2)

Skin tag (757.31)
Naevus (757.38)
Angioma (228.0)
Haemangioma (228.0)
Glomus tumor (228.0)
Lymphangioma (228.1)
Lymphatic naevus (228.1)
Birthmark (757.38)
Mongolian blue spot (757.38)

} Are to be reported only if multiple or giant (> 4 cm^2)

Hypospadias when the meatus lies before the coronary sulcus (part of 752.60) (NB. hypospadias should be registered when the meatus lies in/or behind the coronary sulcus).

<u>The following conditions are recognized to be difficult to diagnose and therefore should not be reported, if occurring as a single malformation, to EUROCAT until the anatomical abnormality is confirmed</u>.

- Cardiac murmurs
- Congenital dislocation of the hip
- Clicking hip
- Varus conditions of the feet (postural origin)
- Valgus conditions of the feet (postural origin).

EUROCAT REPORT on CONGENITAL MALFORMATIONS and TWINS

Centre

CD

Local ID No.

Eurocat No.

INFANT 1

Date of Birth

Place of Birth (specify)

Sex (1 = Male 3 = Indeterminate 2 = Female 9 = Not known)

No. of babies delivered

Type of Birth (1 = live 3 = spont. ab. 9 = NK 2 = still 4 = induced ab.)

Birth weight (grams)

Date of L.M.P.

Length of gestation (weeks)

Certainty of L.M.P. (1 = certain 3 = no L.M.P. 2 = uncertain 9 = not known)

Pre-natal diagnosis (1 = result known 3 = not done 2 = result unknown 9 = not known)

Karyotype (code as for box 40) _____

Post mortem examination (code as for box 40)

Source of information (see instructions)

MULTIPLE BIRTHS (see coding instructions)

Birth order (in multiple set)

No. of malformed babies / fetuses

Zygosity determination (if available)

Sex

Chorionic membrane

Amniotic membrane

Other (specify) _____

Name _____

Address _____

_____ Post Code _____

(For local use only)

MOTHER 2

Residence code

Date of Birth

Age at delivery (years)

Previous Pregnancies :- Spontaneous abortion(s) no.

Induced abortion(s) no.

Live births - no.

Stillbirths - no.

Total Pregnancies - no.

Occupation _____

Social status

Racial type _____

For 42-49 code: 1 = yes; 2 = no; 9 = NK :-

Previous children with anomalies _____

Family history of anomaly (state relationship and anomaly) _____

Chronic illness (specify) _____

Illness during pregnancy (specify) _____

Smoking (no. per day) _____

Alcohol (amount per week) _____

Drugs (specify) _____

Family history of twinning

Consanguinity (1 = 1st cousin 8 = Other relation 9 = NK 2 = 2nd cousin 0 = Not related)

Enter additional malformations / comments below:-

FATHER

Date of Birth

Age at delivery (years)

Occupation _____

Social status

Racial type _____

For 66 -71 code: 1 = yes; 2 = no; 9 = NK:-

Family history of anomaly (state relationship and anomaly) _____

Chronic illness (specify) _____

Smoking (no. per day) _____

Alcohol (amount per week) _____

Drugs (specify) _____

Family history of twinning

MALFORMATIONS 3

Date of discovery

When discovered (see instructions)

Condition at discovery (1 = live 2 = dead 9 = NK)

Syndrome _____

Malformations present:

1 _____

2 _____

3 _____

4 _____

5 _____

6 _____

7 _____

8 _____

Confirmation of diagnosis (see coding instructions)

Mode of inheritance (see coding instructions)

No. of malformations (to be coded centrally)

General instructions: One form is to be completed for each abnormal baby or fetus.

One form is to be completed for each baby or fetus from multiple births, except in the case of conjoined twins when only one form should be completed.

In general, the code '8' should be used for 'other', and

the code '9' should be used for 'not known'.

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
				1
Block Ref. 1				
Local ID No.	11 (max.)	Use local code	In the interests of confidentiality, this information will not be computerised by EUROCAT	-
EUROCAT No.	11	**Registry code number** 01 : W. Vlaanderen (B) 02 : Hainaut (B) 03 : Odense (DK) 04 : Morlaix (F) 05 : Yvelines et Paris (F) 06 : Frankfurt (D) 07 : W. Berlin (D) 08 : Firenze (I) 09 : Roma (I) 10 : Dublin (IRL) 11 : Galway (IRL) 12 : Luxembourg (L) 13 : Groningen (NL) 14 : G Glasgow (UK) 15 : Liverpool (UK) 16 : Belfast (UK) 17 : Eubia (GR) 18 : 19: 20:	Enter the code allocated to you in the first two boxes. The remaining boxes will be completed by EUROCAT -	1-2 3-11
Block reference	1	1		12
Date of Birth	6		Day / Month / Year e.g. 21st July 1980 code 2 1 0 7 8 0	13-18

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Place of birth	5 (max.)	1 : Public hospital (State financed) 2 : University (Teaching) hospital 3 : Private hospital 4 : G.P. Unit (Family doctor/ practitioner) 5 : Home delivery 7 : Born before admission (BBA) - (on way to hospital) 8 : Other 9 : Not known	Use one of these codes for the <u>first</u> digit to indicate the type of institution, etc. The remaining four digits are for your local code which identifies a specific unit. <u>Do not</u> send the key to EUROCAT.	19 [1] 20-23
Sex	1	1 : Male 2 : Female 3 : Indeterminate 9 : Not known		24
Number of babies / fetuses delivered	1	1 : Singleton 2 : Twins 3 : Triplets .... etc. . . 9 : Not known	If multiple delivery complete boxes 48 - 53 Complete one form for each baby whether normal or malformed. Only <u>one</u> form to be completed for <u>conjoined</u> twins.	25
Type of birth	1	1 : Live birth 2 : Stillbirth 3 : Spontaneous abortion 4 : Induced abortion 9 : Not known	See definitions - Appendix I	26
Birth weight	4	9999 : Not known	Complete the weight in grams. See Appendix I	27-30

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Date of L.M.P. (last menstrual period)	6	999999 : not known 000000 : no L.M.P.	Day / Month / Year e.g. 21st July 1980 2 1 0 7 8 0	31-36 1
Length of gestation	2	00 : impossible to ascertain	In completed weeks. If L.M.P. is available and certain, calculate gestational age from date of L.M.P. to date of delivery. (Appendix I) If L.M.P. is available but uncertain, give the corrected gestational age by clinical ascertainment or other means. If L.M.P. is not available, give the estimated gestational age by clinical ascertainment or other means.	37-38
Certainty of L.M.P.	1	1 : certain 2 : uncertain 0 or 3 : no LMP 9 : not known		39
Pre-natal diagnosis	1	1 : performed, result known 2 : performed, result unknown 9 : not known 0 or 3 : not performed	If performed, record indication(s), investigation(s), and results in the space for comments etc. or on the back.	40
Karyotype	1	1 : performed, result known 2 : performed, result unknown 9 : not known 0 or 3 : not performed	If performed, record the results in the 'comments' space or on the back of the form.	41

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Post mortem examination	1	1 : performed, result known 2 : performed, result unknown 9 : not known 0 or 3 : not performed	If performed record the malformation(s) discovered in the 'Malformations' section of the form. If other findings, record on the back of the form or in the 'comments' space.	42 [1]
Source of information	5 (max.)	1 : Notes in routine scan 2 : Birth notification or notification of malformation at birth 3 : Hospital case notes 4 : Death or stillbirth certificate 5 : Pre-natal diagnosis report 6 : Laboratory report (cytogenetic, chemical, etc.) 7 : Post mortem examination 8 : Other 9 : Not known	Indicate each source of information - up to a maximum of five - used in collecting information on the child.	43-47
MULTIPLE BIRTHS			This section only to be completed for multiple births	48-53
Birth order	1	1, 2, 3 etc 9 : multiple birth, order unknown		48
Number of malformed babies/fetuses	1	1, 2, 3 etc 9 : multiple birth, number unknown		49
Zygosity determination Sex	1	1 : like sex 2 : unlike sex 3 : indeterminate sex 8 : other 9 : multiple, sex not stated	If more than two fetuses / infants of unlike sex use code '8'.	50

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Chorionic membrane	1	1 : monochorial 2 : dichorial 8 : other 9 : if multiple and not stated	See EUROCAT booklet	51 [1]
Amniotic membrane	1	1 : monoamniotic 2 : diamniotic 8 : other 9 : if multiple and not stated	See EUROCAT booklet	52
Other means of zygosity determination	1	1 : yes 2 : no 9 : not known	If method other than above used, specify on form and code '1' for yes.	53

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Block Ref. 2 MOTHER	1	2		12 [2]
Residence code	7 (max.)		Use local code for locality (area) of residence. Send key code to EUROCAT	13-19
Date of birth	6	999999 : if not stated	Day / Month / Year e.g. 1st October 1945 code [0][1][1][0][4][5]	20-25
Age at delivery	2	99 : if not known	In completed years at the time of delivery	26-27
Number of spontaneous abortions	1	9 : if not known	If a twin pregnancy aborted, code as 2 See definitions - Appendix I	28
Number of induced abortions	1	9 : if not known	As for box 28	29
Number of live births	2	99 : if not known	A twin delivery counts as 2 if both live born, triplets as 3, etc.	30-31
Number of stillbirths	1	9 : if not known	A twin delivery counts as 2 if both stillborn, triplets as 3, etc.	32
Total number of previous pregnancies	2	99 : if not known	This is total previous pregnancies not births. The present notified pregnancy is not to be included. Include miscarriages (abortions). Twin or other multiple pregnancies count as 1 in the total. e.g. 3 previous LB + 1 termination of a twin pregnancy = [0][4]	33-34

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Occupation	5		Give as much detail as possible. Give main occupation at the time of conception. Give job, industry, status in verbal description, if possible (English/French). To be coded locally. Send source of code to EUROCAT	35-39 2
Social status	1	Leave blank	This will be coded locally from occupation	40
Racial type	1		Use local code as used for local birth or census data. Send key of code to EUROCAT	41
Previous child(ren) with anomaly(ies)	1	1 : yes 0 or 2 : no 9 : not known	Specify if 'yes'	42
Family history of anomaly(ies)	1	1 : yes 0 or 2 : none 9 : not known	As for 42. Also, state relationship.	43
Chronic illness	1	1 : yes 0 or 2 : no 9 : not known	As for 42.	44
Illness during pregnancy (acute)	1	1 : yes 0 or 2 : no 9 : not known	As for 42.	45
Smoking	1	1 : yes 0 or 2 : no 9 : not known	If yes - give no. per day	46

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Alcohol	1	1 : yes 0 or 2 : no 9 : not known	If yes - give amount and type per day	47 [2]
Drugs during pregnancy	1	1 : yes 0 or 2 : no 9 : not known	If yes - give as much detail as possible	48
Family history of twinning	1	1 : yes 0 or 2 : no 9 : not known		49
Consanguinity	1	1 : 1st cousin 2 : 2nd cousin 8 : other relation or relation not stated 9 : not known 0 : not related		50
FATHER			See same items under 'Mother' and use the same codes	51-71

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
MALFORMATIONS				3
Block Ref. 3	1			12
Date of discovery	6	999999 : not stated	The date on which the baby was first suspected or recognized as being malformed, even if the detailed diagnosis is not available. Day / Month / Year e.g. 14th January 1980 code 1 4 0 1 8 0	13-18
When discovered	1	1 : birth 2 : less than 1 week 3 : 1 - 4 weeks 4 : 1 - 12 months 5 : over 12 months 6 : pre-natal diagnosis 7 : spontaneous or induced abortion 9 : not known		19
Condition at discovery	1	1 : live 2 : dead 9 : not known		20
Syndrome, eponym or disease name	6	ICD 9th revision code or modifications stated in next column	Use ICD 9th revision code in the first four digits, the British Paediatric Association Classification of Diseases in the fifth digit, and the EUROCAT Guide to Eponyms and Syndromes in the sixth digit. If not a recognizable syndrome, eponym, or disease name leave blank. If a recognizable syndrome, eponym or disease name with no ICD code, then code 888888 and notify Dr. Weatherall (see forward to EUROCAT Guide to Eponyms and Syndromes).	21-26

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Syndrome, ... etc. contd.			It is recommended that all the abnormalities observed by the local clinician should be coded in addition to the specific name. For the written descriptions, please use English or French. If only the name of the specific syndrome, eponym or disease is available, code in boxes 21-26 and leave the remainder blank (i.e. boxes 27-74). e.g. Trisomy 18 / Edwards syndrome, code 7 5 8 2 0 0 in boxes 21-26 When two syndromes are present in the same subject, code the more important in 27-32.	contd. 3
31 Malformations present	6	ICD 9th revision code or modifications already stated	When a specific syndrome, eponym or disease is reported in 21-26 list the observed abnormalities from box 27 on. These abnormalities should be coded as if they are isolated abnormalities in the ICD e.g. congenital rubella embryopathy with bilateral cataracts and unspecified abnormality of heart code as 7 6 0 2 [ ] [ ] in 21-26 and in boxes 27-32 bilateral cataracts 7 4 3 3 2 [ ] and in 33-38 unspecified heart abnormality 7 4 6 9 9 [ ] Do not indicate multiple minor abnormalities which are part of a well established named specific syndrome, eponym or disease, e.g. Down's syndrome with a ventricular septal defect and Brushfield spots. Enter Down's syndrome 7 5 8 0 9 [ ] in 21-26 and enter ventricular septal defect 7 4 5 1 4 [ ] in 27-32, but omit Brushfield spots.	27-74

ITEM	DIGITS	CODE	INSTRUCTIONS & NOTES	BOX NO.
Confirmation of diagnosis	1	1 : confirmed 2 : still doubtful 3 : lost to follow-up 9 : not known 0 : no malformation	In cases for which the diagnosis of malformation is well established - transmit monthly, together with those which are certainly malformed but for which the exact diagnosis is not yet confirmed. When this is confirmed or further information is available an amended report should be sent. In cases where a malformation is only suspected these should be recorded locally and sent to EUROCAT only when the diagnosis is confirmed. If after a year the diagnosis has not been confirmed or the case has been lost to follow-up the forms should then be sent to EUROCAT as a separate batch.	75 [3]
Mode of inheritance	1	0 : autosomal dominant 1 : autosomal recessive 2 : X-linked recessive / dominant 3 : numeric chromosomal defect of autosomes 4 : structural chromosomal defect of autosomes 5 : numeric sex chromosomal defects 6 : structural sex chromosomal defects 7 : polygenic / multifactorial 8 : known environmental agent 9 : other or unknown	Use OPCS recommendations	76
Number of malformations	2	Leave blank	This will be coded centrally	77-78

APPENDIX VII : DEFINITIONS

1. Live birth

Live birth is the complete expulsion or extraction from its mother of a product of conception, irrespective of the duration of the pregnancy, which, after such separation, breathes or shows any other evidence of life, such as beating of the heart, pulsation of the umbilical cord, or definite movement of voluntary muscles, whether or not the umbilical cord has been cut or the placenta is attached; each product of such a birth is considered live born. (WHO definition)

2. Stillbirth

Stillbirth is the complete expulsion or extraction from its mother of a product of conception of 28 weeks (196 days) gestation or more, which shows no evidence of life such as breathing, spontaneously or after stimulation, beating of the heart, or pulsation of the umbilical cord, or when post mortem examination fails to show air expansion in the lungs. (If your definition differs from above, please send a copy to EUROCAT.)

3. Abortion

Abortion is the expulsion, complete or incomplete, or extraction of the products of conception prior to 28 completed weeks of pregnancy (up to 195 days) which show no evidence of life such as breathing spontaneously, beating of the heart, pulsation of the umbilical cord, or definite movement of voluntary muscles. (If your definition differs from above, please send a copy to EUROCAT.)

4. Birthweight

The first weight of the fetus or newborn obtained after birth. This weight should be measured preferably within the first hour of life before significant postnatal weight loss has occurred. (WHO definition)

5. Gestational age

The duration of gestation is measured from the first day of the last normal menstrual period. Gestational age is expressed in completed days or completed weeks (e.g. events occurring 280 to 286 days after the onset of the last normal menstrual period are considered to have occurred at 40 weeks of gestation). (WHO definition)