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The baseline data of the Hungarian Congenital Malformation Register, 1970–1976

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The baseline data of the Hungarian Congenital Malformation Register 1970—1976 are published for national and international use and evaluation

The renewed Hungarian Congenital Malformation Register (HCMR) was launched January 1st, 1970. Its aim and function were described earlier [1]. The classification of categories and subcategories of congenital anomalies is based on the International Classification of Diseases (ICD), VIIIth Revision, 1965 [3], mainly on its Chapter XIV entitled "Congenital Anomalies". But the HCMR shows some deviations from the ICD:

(i) All *notified* congenital anomalies, e.g. congenital rubella syndrome, inborn errors of metabolism, congenital hernias and tumors, etc., have been included.

(ii) A new type of subcategory has been established, of the so-called "*complex*" congenital malformation which involves two or more notified congenital malformations within the *same* organ, e.g. heart, eye, bowels, etc.

Table I

Number and occurrence of malformed babies in Hungary, 1970—1976

Year	Live births		Stillbirths		Total births		Number of babies with congenital malformations	Occurrence of malformed babies per 1000 total births
	No.	%	No.	%	No.	%		
1970	151,819	99.01	1520	0.99	153,339	100.00	3304	21.55
1971	150,640	99.00	1519	1.00	152,159	100.00	4355	28.62
1972	153,265	99.10	1423	0.90	154,688	100.00	4802	31.04
1973	156,224	99.11	1399	0.89	157,623	100.00	4780	30.33
1974	186,288	99.11	1669	0.89	187,957	100.00	6301	33.52
1975	194,240	99.18	1607	0.82	195,847	100.00	6909	35.53
1976	185,395	99.19	1521	0.81	186,916	100.00	6844	36.56

TABLE II

Number and occurrence of registered congenitally malformed newborn infants according to categories and subcategories of ICD VIIIth revision in the Hungarian Congenital Malformations Register, 1970—1976

ICD code number	Categories and subcategories	1970		1971		1972		1973		1974		1975		1976	
		No.	‰	No.	‰	No.	‰	No.	‰	No.	‰	No.	‰	No.	‰
740.	Anencephalus	145	0.95	130	0.85	156	1.00	117	0.74	181	0.96	128	0.66	118	0.63
741.	Spina bifida	189	1.23	184	1.21	164	1.05	146	0.93	192	1.02	156	0.81	140	0.75
742.	Congenital hydrocephalus	48+	0.31	98	0.64	93	0.60	100	0.63	148	0.79	149	0.77	114	0.61
743.0	Encephalocele	38	0.25	37	0.24	48	0.31	37	0.24	44	0.23	31	0.16	31	0.16
743.1	Microcephalus	12	0.08	23	0.15	22	0.15	10	0.06	28	0.15	29	0.14	22	0.12
743.2**—743.9	Other anomalies of nervous system	2	0.01	7	0.05	16	0.11	12	0.08	19	0.10	17	0.09	12	0.06
744.0—744.1	Anophthalmos and microphthalmos	4	0.03	11	0.07	5	0.03	1	0.01	3	0.02	0	—	3	0.20
744.3 744.2	Congenital cataract	10	0.07	8	0.05	5	0.03	3	0.02	9	0.05	3	0.01	9	0.05
744.4—744.9	Other anomalies of eye	13	0.08	24	0.16	22	0.15	28	0.18	21	0.11	23	0.11	15	0.08
745.0 745.2	Anomalies of ear	18	0.12	20	0.13	19	0.12	17	0.11	24	0.13	33	0.17	30	0.16
745.1	Accessory auricle	11	0.07	11	0.07	8	0.05	6	0.04	10	0.05	7	0.03	11	0.06
745.4	Branchial cleft cyst, or fistula; preauricular sinus	8	0.05	10	0.07	9	0.06	10	0.06	6	0.03	13	0.07	6	0.03
745.7* 745.5	Torticollis	1	0.01	6	0.04	48	0.31	31	0.20	30	0.16	23	0.11	7	0.04
745.8—745.9	Other anomalies of face and neck	4	0.03	6	0.04	7	0.05	3	0.02	5	0.03	7	0.03	9	0.05
746.0	Common truncus	6	0.04	6	0.04	10	0.06	6	0.04	13	0.07	13	0.07	9	0.05
746.1	Transposition of great vessels	11	0.07	19	0.12	17	0.12	18	0.11	38	0.20	31	0.17	30	0.16
746.2	Tetralogy of Fallot	16	0.10	26	0.17	14	0.09	17	0.11	19	0.10	18	0.09	22	0.12
746.3	Ventricular septal defect	94	0.61	119	0.78	162	1.05	148	0.94	159	0.85	181	0.93	170	0.91
746.4	Atrial septal defect	37	0.24	33	0.22	31	0.20	37	0.24	41	0.22	43	0.22	40	0.21
746.5—746.8	Other specified anomalies of heart	9	0.06	11	0.07	23	0.15	38	0.24	56	0.30	35	0.18	27	0.14
747.7*	Complex congenital anomalies of heart	33	0.22	48	0.31	44	0.28	63	0.40	67	0.36	73	0.38	61	0.33

746.9	Unspecified anomalies of heart	116	0.76	182	1.20	166	1.07	170	1.08	237	1.25	348	1.78	307	1.65
747.0	Patent ductus arteriosus	18	0.12	14	0.09	31	0.22	33	0.21	20	0.11	36	0.18	53	0.28
747.1	Coarctation of aorta	1	0.01	4	0.03	6	0.04	7	0.04	6	0.03	13	0.07	9	0.05
747.2	Other anomalies of aorta	4	0.03	21	0.14	18	0.12	15	0.10	24	0.13	30	0.15	21	0.11
747.3	Stenosis or atresia of pulmonary artery	3	0.02	8	0.05	0	—	11	0.07	15	0.08	15	0.07	15	0.08
747.4—747.6	Other anomalies of circulatory system	1	0.01	2	0.01	3	0.02	7	0.04	7	0.04	10	0.05	6	0.03
747.8—747.8															
748.	Congenital anomalies of respiratory system	13	0.08	25	0.16	29	0.19	26	0.17	28	0.15	31	0.16	29	0.16
749.0	Cleft palate	44	0.29	49	0.32	43	0.28	45	0.29	60	0.32	58	0.30	63	0.33
749.1—749.2	Cleft lip; cleft palate with cleft lip	123	0.80	145	0.95	163	1.05	149	0.95	171	0.91	169	0.87	171	0.91
749.3*	Robin anomalad	5	0.03	7	0.05	6	0.04	8	0.05	14	0.07	16	0.08	9	0.05
750.1	Pyloric stenosis	44	0.29	87	0.57	58	0.37	44	0.28	57	0.30	*48*	0.25	57	0.30
750.2	Tracheo-oesophageal fistula, oesophageal atresia and stenosis	19	0.12	27	0.18	30	0.19	19	0.12	32	0.17	32	0.16	40	0.21
751.1	Atresia and stenosis of small intestine	7	0.05	30	0.20	28	0.18	27	0.17	24	0.13	34	0.17	43	0.23
751.2	Atresia and stenosis of rectum and anal canal	24	0.16	32	0.21	16	0.11	35	0.22	33	0.18	39	0.20	31	0.16
751.2	Hirschsprung disease	5	0.03	11	0.07	23	0.15	16	0.10	10	0.05	17	0.09	13	0.07
751.6—750.0**750.8—751.0	Anomalies of gallbladder, bile ducts, liver and pancreas	8	0.05	13	0.08	13	0.09	14	0.09	22	0.12	22	0.11	21	0.11
751.4—751.5751.8—751.9	Other anomalies of digestive system	5	0.03	22	0.14	18	0.12	17	0.11	43	0.23	21	0.11	29	0.16
752.1	Undescended testicle	26	0.17	38	0.25	90	0.58	88	0.56	110	0.59	169	0.87	168	0.90
752.2—752.3	Hypospadias and epispadias	77	0.50	164	1.08	153	0.99	200	1.27	235	1.25	303	1.55	336	1.80
752.4	Congenital hydrocele	77	0.50	93	0.61	93	0.60	78	0.49	156	0.83	169	0.87	239	1.28
752.0**752.5—752.9	Other anomalies of genital organs	26	0.17	12	0.08	18	0.12	17	0.11	16	0.09	21	0.11	19	

TABLE II (Cont.)

ICD code number	Categories and subcategories	1970		1971		1972		1973		1974		1975		1976+	
		No.	‰	No.	‰	No.	‰	No.	‰	No.	‰	No.	‰	No.	‰
753.0**	Renal agenesis	4	0.03	12	0.08	7	0.05	8	0.05	11	0.06	4	0.02	13	0.07
753.01*	Caudal regression anomalad	1	0.01	4	0.03	8	0.05	15	0.10	9	0.05	10	0.05	20	0.11
753.02*	Triad anomalad	1	0.01	0	—	2	0.01	1	0.01	5	0.03	4	0.02	5	0.03
753.1	Cystic kidney disease	3	0.02	10	0.07	21	0.14	16	0.10	11	0.06	21	0.11	15	0.08
753.2	Obstructive defects of urinary tract	0	—	3	0.02	11	0.07	19	0.12	28	0.15	17	0.09	17	0.09
753.5	Exstrophy of urinary bladder and cloacae	5	0.03	0	—	5	0.03	8	0.05	8	0.04	12	0.06	9	0.05
753.3—753.4 753.8—753.9	Other anomalies of urinary system	8	0.05	22	0.14	23	0.15	19	0.12	26	0.14	38	0.19	26	0.14
754.0	Clubfoot (congenital)	170	1.11	239	1.57	247	1.60	258	1.64	326	1.73	378	1.94	337	1.81
755.0	Polydactyly	46	0.30	85	0.55	97	0.63	71	0.45	123	0.65	91	0.47	98	0.53
755.01*	Polydactily + syndactyly	7	0.05	4	0.03	8	0.05	8	0.05	13	0.07	11	0.05	18	0.10
755.1	Syndactyly	19	0.12	38	0.25	46	0.30	34	0.22	44	0.23	67	0.34	43	0.23
755.2—755.4	Reduction deformity of limbs	28	0.18	44	0.29	37	0.24	45	0.29	63	0.34	89	0.46	82	0.44
755.6 755.5	Congenital dislocation of hip	572	3.73	722	4.75	809	5.23	854	5.42	1142	6.08	1422	7.26	1514	8.10
755.7—755.9	Other anomalies of limbs	12	0.08	7	0.05	16	0.11	14	0.09	9	0.05	18	0.09	15	0.08
756.**	Other congenital anomalies of musculoskeletal system	20	0.13	22	0.14	15	0.10	23	0.15	30	0.16	25	0.13	23	0.12
757.1	Pigmented naevus	37	0.24	20	0.13	44	0.28	35	0.22	52	0.28	74	0.38	102	0.55
757.0 757.2—575.9	Other anomalies of skin, hair and nails	3	0.02	11	0.07	15	0.10	8	0.05	13	0.07	18	0.09	5	0.03
758.2	Anomalies of thyroid gland	6	0.04	8	0.05	15	0.10	24	0.15	34	0.18	16	0.08	15	0.08
758.0—758.1 758.3	Anomalies of spleen and other endocrine glands	1	0.01	2	0.01	0	—	5	0.03	5	0.03	9	0.04	2	0.01
758.9	Unspecified congenital anomaly	0	—	5	0.03	0	—	5	0.03	3	0.02	9	0.04	0	—
759.0	Situs inversus	5	0.03	2	0.01	2	0.01	2	0.01	5	0.03	15	0.07	2	0.01
759.1*	Conjoined twins	2	0.01	2	0.01	2	0.01	1	0.01	7	0.04	1	0.00	5	0.03

759.2*	Relatively frequent heterogeneous multiple malformations														
759.211*	Chondrodystrophia	3	0.02	12	0.08	17	0.12	11	0.07	8	0.04	14	0.07	21	0.11
759.212*	Osteogenesis imperfecta	7	0.05	4	0.03	9	0.06	5	0.03	4	0.02	7	0.03	5	0.03
759.213*	Arthrogryposis	4	0.03	4	0.03	5	0.03	4	0.03	0	—	3	0.01	4	0.02
759.22*	Structural syndromes due to major gene defect	11	0.07	13	0.08	10	0.06	12	0.08	18	0.10	26	0.13	10	0.05
759.23*	Inborn errors of metabolism	6	0.04	9	0.06	15	0.10	12	0.08	24	0.13	33	0.17	28	0.14
759.23*	Heredoneuromuscular anomalies	1	0.01	3	0.02	1	0.01	3	0.02	4	0.02	4	0.02	8	0.04
759.3	Down disease	87	0.57	111	0.73	128	0.83	136	0.86	175	0.93	168	0.86	169	0.90
759.4	Other syndromes due to autosomal abnormality	7	0.05	4	0.03	12	0.08	13	0.08	10	0.05	10	0.05	12	0.06
759.5	Syndromes due to sex chromosome abnormality	3	0.02	3	0.02	5	0.03	3	0.02	5	0.03	5	0.02	6	0.03
759.6*	Structural syndromes due to environmental agents	3	0.02	4	0.03	0	—	4	0.03	15	0.08	11	0.05	10	0.05
759.71*	Postural associations	83	0.55	110	0.72	128	0.83	133	0.85	97	0.52	61	0.31	35	0.21
759.82*	Combinations														
759.821*	Persons with two malformations														
	Cardinal malformations														
759.8211*	First group														
	Anencephalus	8	0.05	9	0.06	16	0.11	22	0.14	18	0.10	18	0.09	16	0.08
	Encephalocele	5	0.03	6	0.04	3	0.02	6	0.04	11	0.06	9	0.04	1	0.01
	Spina bifida	14	0.09	17	0.11	18	0.12	20	0.13	31	0.16	26	0.13	19	0.10
	Cleft lip; cleft lip with cleft palate	13	0.08	14	0.09	19	0.12	30	0.19	11	0.06	22	0.11	24	0.13
	Cleft palate	6	0.04	10	0.07	10	0.06	9	0.06	9	0.05	6	0.03	10	0.05
	Reduction deformity of limbs	5	0.03	3	0.02	6	0.04	13	0.08	10	0.05	7	0.03	9	0.05
	Polydactyly	3	0.02	8	0.05	14	0.09	8	0.05	9	0.05	12	0.06	10	0.05
	Syndactyly	7	0.05	9	0.06	4	0.03	6	0.04	13	0.07	17	0.09	8	0.04
	Exomphalos	2	0.01	8	0.05	6	0.04	9	0.06	3	0.02	7	0.03	9	0.05
	Oesophageal atresia	8	0.05	5	0.03	5	0.03	7	0.04	6	0.03	8	0.04	3	0.02
	Atresia of anal canal	8	0.05	9	0.06	9	0.06	7	0.04	4	0.02	7	0.03	9	0.05
759.8212*	Second group														
	Microcephalus	8	0.05	5	0.03	7	0.05	8	0.05	6	0.03	12	0.06	9	0.05
	Hydrocephalus	7	0.05	13	0.08	16	0.11	18	0.11	15	0.08	14	0.07	16	0.08
	Anomalies of eye	4	0.03	5	0.03	7	0.05	8	0.05	3	0.02	6	0.03	1	0.01
	Clubfoot	6	0.04	10	0.07	10	0.06	6	0.04	9	0.05	6	0.03	21	0.11
	Congenital dislocation of hip	6	0.04	7	0.05	19	0.12	15	0.09	26	0.14	26	0.13	34	0.18
	Hypospadias	12	0.08	10	0.07	17	0.12	12	0.08	22	0.12	27	0.14	23	0.12

TABLE II (Cont.)

ICD code number	Categories and subcategories	1970		1971		1972		1973		1974		1975		1976+	
		No.	‰	No.	‰	No.	‰	No.	‰	No.	‰	No.	‰	No.	‰
	Undescended testicles	5	0.03	3	0.02	11	0.07	5	0.03	9	0.05	10	0.05	11	0.06
	Diaphragmatic hernia	5	0.03	5	0.03	4	0.03	5	0.03	4	0.02	6	0.03	7	0.04
	Anomalies of heart	19	0.12	29	0.19	43	0.28	40	0.25	36	0.19	43	0.22	48	0.26
	Anomalies of urinary system	0	—	1	0.01	3	0.02	5	0.03	10	0.05	6	0.03	2	0.01
	Hernias	11	0.07	11	0.07	14	0.09	11	0.07	10	0.05	12	0.06	18	0.10
	Anomalies of skin	7	0.05	8	0.05	4	0.03	6	0.04	4	0.02	6	0.03	3	0.02
	Other anomalies	1	0.01	0	—	1	0.01	1	0.01	0	—	0	—	0	—
759.822*	Persons with three or more malformations														
759.8221*	First group														
	Anencephalus	3	0.02	7	0.05	3	0.02	6	0.04	6	0.03	9	0.04	9	0.05
	Encephalocele	1	0.01	3	0.02	1	0.01	4	0.03	0	—	4	0.02	5	0.03
	Spina bifida	7	0.05	5	0.03	8	0.05	11	0.07	13	0.07	10	0.05	8	0.04
	Cleft lip; cleft lip with cleft palate	4	0.03	9	0.06	16	0.11	9	0.05	21	0.11	22	0.11	18	0.10
	Cleft palate	5	0.03	10	0.07	9	0.06	2	0.01	6	0.03	7	0.03	14	0.07
	Reduction deformity of limbs	1	0.01	3	0.02	5	0.03	6	0.04	6	0.03	7	0.03	6	0.03
	Polydactyly	4	0.03	2	0.01	7	0.05	10	0.06	10	0.05	7	0.03	6	0.03
	Syndactyly	3	0.02	3	0.02	3	0.02	3	0.02	4	0.02	6	0.03	0	—
	Exomphalos	0	—	2	0.01	4	0.03	3	0.02	2	0.01	5	0.02	0	—
	Oesophageal atresia	1	0.01	1	0.01	4	0.03	5	0.03	5	0.03	7	0.03	4	0.02
	Atresia of anal canal	1	0.01	4	0.03	5	0.03	3	0.02	6	0.03	7	0.03	3	0.20
759.8222*	Second group														
	Microcephalus	2	0.01	6	0.04	2	0.01	3	0.02	6	0.03	7	0.03	1	0.01
	Hydrocephalus	1	0.01	2	0.01	5	0.03	7	0.04	4	0.02	5	0.02	4	0.02
	Anomalies of eye	0	—	1	0.01	0	—	1	0.01	2	0.01	0	—	3	0.02
	Clubfoot	3	0.02	7	0.05	6	0.04	7	0.04	5	0.03	10	0.05	10	0.05
	Congenital dislocation of hip	2	—	1	0.01	1	0.01	1	0.01	2	0.01	3	0.01	5	0.03
	Hypospadias	0	—	3	0.02	2	0.01	1	0.01	3	0.02	2	0.01	7	0.04
	Undescended testicles	0	—	1	0.01	1	0.01	2	0.01	2	0.01	2	0.01	1	0.01
	Diaphragmatic hernia	1	0.01	0	—	2	0.01	0	—	3	0.02	0	—	4	0.02
	Anomalies of heart	5	0.03	3	0.02	7	0.05	2	0.01	5	0.03	4	0.02	8	0.40
	Anomalies of urinary system	0	—	0	—	0	—	0	—	0	—	0	—	2	0.01
	Hernias	0	—	0	—	2	0.01	0	—	0	—	0	—	1	0.01
	Anomalies of skin	0	—	0	—	0	—	0	—	0	—	0	—	0	—

	Other malformations	0	—	0	—	0	—	0	—	1	0.01	0	—	0	—
759.9	Multiple congenital anomalies, unspecified	27	0.18	20	0.13	25	0.16	38	0.24	57	0.30	5	0.02	5	0.09
550	Inguinal hernia	183	1.19	206	1.35	259	1.67	211	1.34	463	2.46	552	2.84	627	3.35
550.1*	Ing. + umb. hernia	12	0.08	12	0.08	3	0.02	5	0.03	13	0.07	18	0.09	9	0.05
551.11	Omphalocele, exomphalos	19	0.12	20	0.13	29	0.19	28	0.18	33	0.17	39	0.19	49	0.26
551.12	Umbilical hernia	81	0.53	87	0.57	56	0.36	49	0.31	73	0.39	87	0.45	104	0.56
551.3	Diaphragmatic hernia	12	0.08	23	0.15	28	0.20	41	0.26	53	0.28	37	0.18	33	0.18
551.0 551.2 551.8— 551.9	Other hernia of abdominal cavity	4	0.03	6	0.04	9	0.06	8	0.05	10	0.05	9	0.04	18	0.10
227.	Haemangioma	224	1.46	329	2.16	337	2.18	336	2.13	431	2.29	388	1.98	318	1.70
228.	Teratoma	8	0.05	13	0.08	9	0.06	16	0.10	24	0.13	27	0.14	12	0.06
	Grand total No.	3304		4356		4802		4778		6308		6904		6844	
	%	21.55		28.63		31.04		30.31		33.56		35.25		36.62	

* new subcategory established in the HCMR
** syndromes are excluded and they are within the three digit category 759

(iii) An attempt has been made to adopt some new international recommendations for the classification of congenital malformations, e.g. *anomalads* [2].

(iv) In our definition, multiple malformation means *two* or more major malformations in different organs or tissues of a person without direct relation of cause and effect. Thus, there is a possibility to exclude the confusion caused by two different parameter systems: congenital anomalies and malformed babies.

(v) Regarding multiple malformations (three digit category 759) a new classification has been developed in order to establish an easier and more efficacious evaluation.

This paper contains only the data of the HCMR, 1970—1976, without any evaluation and discussion. Its only purpose is to publish the baseline data of the HCMR for Hungarian and international use and evaluation before the HCMR will be transformed from the VIIIth Revision of ICD into the IXth Revision and before a new computer programme will be started on January 1st, 1978.

All occurrences of the different categories and subcategories of congenitally malformed babies (the so-called point-prevalences at birth) are calculated per 1000 total births. The figures for live births, stillbirths and total births in Hungary, 1970—1976, are to be found in Table I.

Table II shows only the more frequent (with occurrences over 1 : 30,000 total births) and well-defined categories and subcategories of congenital anomalies. The rare and inaccurately defined or unspecified congenital anomalies within each three digit code number are collected into joint categories.

The results of statistical utilization of malformed babies according to the month of birth; residence of parents (mother); sex ratio; rate of live births and still births; infant death; singletons and multiple births; birth weight and other parameters will be published later in detail.

These baseline figures are considered to be the final version of the HCMR, 1970—1976. The figures deviate somewhat from the earlier Hungarian publications ("Annual Report of the HCMR") due to some changes in the classifications and modifications made after the deadline.

References

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