Incidence of Congenital Heart Defects in the Czech Republic

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Summary:

Aim of study: An analysis of incidence of congenital heart defects (CHD) in the Czech Republic in the 1994 – 2008 period. An assessment of absolute numbers, frequencies and incidences for particular selected diagnoses according to 10th decennial revision of International Classification of Diseases (ICD-10). An analysis of pre- and postnatal incidences of selected diagnoses and of a prenatal diagnostics efficiency in the Czech Republic.

Type of study: A retrospective epidemiological analysis of congenital anomalies from the database of the National Register of Birth Defects (NRBD) of the Czech Republic.

Material and methods: Data from the NRBD from the 1994 – 2008 period were used. In our study, CHD incidences (ICD-10 Q20-Q28 Congenital malformations of the circulatory system group) in the Czech Republic were analyzed. First, CHD incidences in births were assessed – absolute numbers, frequencies and incidences for particular selected diagnoses. Second, absolute numbers, frequencies and incidences of particular selected diagnoses in prenatally diagnosed fetuses and a secondary prevention measures efficiency in selected CHD were evaluated. In a third part, survival of babies with CHD during the first year of their life was analyzed.

Results: In the period under the study there were a total of 1 472 610 life births in the Czech Republic. Congenital malformations of the circulatory system (Q20-Q28) present more than 40 % of all registered congenital anomalies and are themselves the most frequent birth defect group in births in the Czech Republic. As a whole 29 133 CHD were diagnosed (197.83 per 10 000 live births) in 18 811 children (127.53 per 10 000 live births) in this period which presents more than 36 % of children born with a congenital anomaly in the Czech Republic during 1994 – 2008. CHD most frequently diagnosed in births were congenital malformations of cardiac septa (total 16 428, 145.05 per 10 000 live births, more than 55 % of all CHD) and congenital malformations of great arteries (total 5 389, 47.58 per 10 000 live births, more than 18 % of all CHD).

Further, prenatally diagnosed CHD were analyzed. Incidences for particular diagnoses as well as percentage of pregnancy termination were assessed. A rate of prenatally diagnosed was 11.35 % in discordant ventriculoarterial connection (Q20.3), 8.35 % in discordant atrioventricular connection (Q20.5), 49.41 % in hypoplastic left heart syndrome (Q23.4), 7.64 % in coarctation of aorta (Q25.1) and 9.71 % in tetralogy of Fallot (Q21.3). These anomalies were parts of chromosomal syndromes in 42.58 % and non-chromosomal syndromes in 9.33 %. There were also associated malformations (from other systems than circulatory one). The most frequent were congenital malformations of the nervous system (Q00-Q07) – 14.59 %, congenital malformations and deformations of the musculoskeletal system (Q65-Q79) – 12.44 %, cleft lip and cleft palate (Q35-Q37) – 7.42 % and congenital malformations of the urinary system (Q60-Q64) – 6.70 %.

In children born with a CHD, 84.53 % were associated with other anomaly, out of which more than 70 % were CHD only. Only about 14 % were associated with anomalies from other (non-circulatory system) groups. Perinatal mortality was highest in hypoplastic left heart syndrome (Q23.4) – 327.1 ‰ and in tetralogy of Fallot (Q21.3) – 6.6 ‰.

Conclusions: The study presents current results of analysis of CHD incidences in the Czech Republic in the 1994 – 2008 period. Children born with a CHD make more than 36 % out of all children born with a congenital anomaly. CHD themselves represents an important part (more than 40 %) of all diagnosed congenital anomalies in the Czech Republic. Over the period of the study there was a slight increase of diagnosed CHD during 1994 – 1999 followed by a slight decrease from 2000 with an exception of 2007 year. The most frequent of diagnosed CHD were ventricular septal defect (Q21.0) and atrial septal defect (Q21.1). Both defects incidences changes influence not only a total CHD but also a total congenital anomalies incidence.

An influence of prenatal diagnostics among the five selected CHD was most important in hypoplastic left heart syndrome (Q23.4), less so in others. In prenatal diagnostics group it is necessary to distinguish between those anomalies which may lead to pregnancy termination (parts of both chromosomal and non-chromosomal syndromes and/or association with other severe anomalies) and those in which pregnancy leads to a delivery (late diagnostics, operabile defects, parental decision).

CHD can be a part of chromosomal syndromes. In our study, in prenatally diagnosed CHD it was more than 42 %. A presence of other associated diagnoses of congenital anomalies in births will significantly influence infant mortality and morbidity.

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Šípek A, Gregor V, Šípek A Jr, Hudáková J, Horáček J, Klaschka J, Skibová J, Langhammer P, Petržílková L, Klímová B, Peřinová B, Wiesnerová J.: Incidence of congenital heart defects in the Czech Republic - current data. Česká Gynekol. 2010 May;75(3):221-42. (in Czech)

Congenital heart defects in the Czech Republic 1994 – 2008 by groups ICD X. (Q20-Q28) - births

ICD-X		n	Incidence	Frequency	95% CI
Q20	Congenital malformations of cardiac chambers and connections	1327	9,01	1110	8,533-9,509
Q21	Congenital malformations of cardiac septa	18823	109,80	91	126,001-129,960
Q22	Congenital malformations of pulmonary and tricuspid valves	2211	15,01	666	14,395-15,653
Q23	Congenital malformations of aortic and mitral valves	2236	15,18	659	14,561-15,827
Q24	Other congenital malformations of heart	1029	6,99	1431	6,567-7,428
Q25	Congenital malformations of great arteries	5289	35,92	278	34,954-36,897
Q26	Congenital malformations of great veins	457	3,10	3222	2,825-3,401
Q27	Other congenital malformations of peripheral vascular system	349	2,37	4220	2,128-2,632
Q28	Other congenital malformations of circulatory system	84	0,57	17531	0,455-0,706

Mortality due congenital heart defects in the Czech Republic 1994 – 2008- selected diagnosis

		Infant mortality rate (per mille)	Stillbirth rate (per mille)	Perinatal mortality rate (per mille)	Neonatal mortality rate (per mille)	Posteneonatal mortality rate (per mille)
Q20.3	Discordant ventriculoarterial connection	65,116	2,320	32,483	58,140	0,017
Q20.5	Discordant atrioventricular connection	78,125	0,000	0,000	31,250	0,756
Q21.3	Tetralogy of Fallot	52,863	6,565	17,505	26,432	0,060
Q23.4	Hypoplastic left heart syndrome	539,906	4,673	327,103	488,263	0,474
Q25.1	Coarctation of aorta	63,988	1,486	26,746	40,179	0,037
Q20.0- Q28.9	Congenital malformations of the circulatory system	38,673	2,020	16,320	24,610	0,001

Congenital heart defects in the Czech Republic 1994 – 2008 – selected diagnosis ICD X. (Q20-Q28)

ICD-X		n	Incidence	Frequency	95% CI
Q200	Common arterial trunk	133	0,90	11072	0,756 - 1,070
Q201	Double outlet right ventricle	268	1,82	5495	1,609 - 2,051
Q202	Double outlet left ventricle	13	0,09	113278	0,047 - 0,151
Q203	Discordant ventriculoarterial connection	445	3,02	3309	2,748 - 3,316
Q204	Double inlet ventricle	83	0,56	17742	0,449 - 0,699
Q205	Discordant atrioventricular connection	65	0,44	22656	0,341 - 0,563
Q208	Other CM of card. chambers and connect.	202	1,37	7290	1,189 - 1,574
Q209	CM of card. chamb. and conn., unspecif.	118	0,80	12480	0,663 – 0,960
Q210	Ventricular septal defect	7813	53,06	188	51,885 – 54,245
Q211	Atrial septal defect	6473	43,96	228	42,892 – 45,040
Q212	Atrioventricular septal defect	498	3,38	2957	3,097 - 3,692
Q213	Tetralogy of Fallot	465	3,16	3167	2,877 - 3,458
Q214	Aortopulmonary septal defect	35	0,24	42075	0,166 - 0,331
Q218	Other CMs of cardiac septa	580	3,94	2539	3,625 – 4,273
Q219	CM of cardiac septum, unspecified	305	2,07	4828	1,845 - 2,317
Q220	Pulmonary valve atresia	146	0,99	10086	0,837 - 1,166
Q221	Congenital pulmonary valve stenosis	711	4,83	2071	4,480 - 5,196
Q222	Congenital pulmonary valve insufficiency	197	1,34	7475	1,157 - 1,538
Q223	Other CMs of pulmonary valve	38	0,26	38753	0,183 - 0,354
Q224	Congenital tricuspid stenosis	56	0,38	26297	0,287 - 0,494
Q225	Ebstein's anomaly	59	0,40	24959	0,305 - 0,517
Q226	Hypoplastic right heart syndrome	38	0,26	38753	0,183 - 0,354
Q228	Other CMs of tricuspid valve	898	6,10	1640	5,706 – 6,510
Q229	CM of tricuspid valve, unspecified	68	0,46	21656	0,359 - 0,585
Q230	Congenital stenosis of aortic valve	531	3,61	2773	3,306 - 3,926
Q231	Congenital insufficiency of aortic valve	421	2,86	3498	2,592 - 3,145
Q232	Congenital mitral stenosis	182	1,24	8091	1,063 - 1,429
Q233	Congenital mitral insufficiency	538	3,65	2737	3,351 – 3,976
Q234	Hypoplastic left heart syndrome	216	1,47	6818	1,278 - 1,676
Q238	Other CMs of aortic and mitral valves	149	1,01	9883	0,856 - 1,188
Q239	CM of aortic and mitral valves, unspecif.	199	1,35	7400	1,170 - 1,553

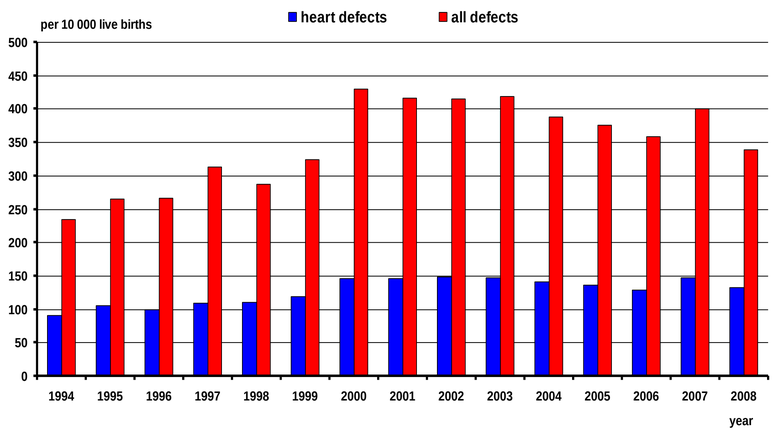
Congenital heart defects in the Czech Republic 1994 – 2008 – selected diagnosis ICD X. (Q20-Q28)

ICD-X		n	Incidence	Frequency	95% CI
Q240	Dextrocardia	113	0,77	13032	0,632 - 0,923
Q241	Laevocardia	8	0,05	184076	0,023 - 0,107
Q242	Cor triatriatum	26	0,18	56639	0,115 - 0,259
Q243	Pulmonary infundibular stenosis	28	0,19	52593	0,126 - 0,275
Q244	Congenital subaortic stenosis	33	0,22	44625	0,154 - 0,315
Q245	Malformation of coronary vessels	141	0,96	10444	0,806 - 1,129
Q246	Congenital heart block	43	0,29	34247	0,211 - 0,393
Q248	Other specified CMs of heart	289	1,96	5096	1,743 - 2,202
Q249	CM of heart, unspecified	348	2,36	4232	2,121 - 2,625
Q250	Patent ductus arteriosus	2861	19,43	515	18,723 – 20,153
Q251	Coarctation of aorta	689	4,68	2137	4,336 – 5,042
Q252	Atresia of aorta	44	0,30	33468	0,217 - 0,401
Q253	Stenosis of aorta	110	0,75	13387	0,614 - 0,900
Q254	Other congenital malformations of aorta	329	2,23	4476	1,999 - 2,489
Q255	Atresia of pulmonary artery	68	0,46	21656	0,359 - 0,585
Q256	Stenosis of pulmonary artery	665	4,52	2214	4,179 – 4,872
Q257	Other CMs of pulmonary artery	206	1,40	7149	1,214 - 1,604
Q258	Other CMs of great arteries	193	1,31	7630	1,132 - 1,509
Q259	CM of great arteries, unspecified	124	0,84	11876	0,700 – 1,004
Q261	Persistent left superior vena cava	182	1,24	8091	01,063 -1,429
Q262	Total anomal. pulm. venous connection	110	0,75	13387	0,614 – 0,900
Q263	Partial anomal. pulm. venous connection	95	0,65	15501	0,522 – 0,789
Q264	Anomal. pulm. ven. connection, unspecif.	19	0,13	77506	0,078 – 0,201
Q268	Other CMs of great veins	27	0,18	54541	0,121 - 0,267
Q269	CM of great vein, unspecified	19	0,13	77506	0,078 – 0,201
Q271	Congenital renal artery stenosis	4	0,03	368153	0,004 – 0,051
Q273	Peripheral arteriovenous malformation	11	0,07	133874	0,007 – 0,070
Q274	Congenital phlebectasia	7	0,05	210373	0,037 – 0,134
Q278	Other specif. CMs of periph. vasc. system	61	0,41	24141	0,317 – 0,532
Q279	CM of periph. vascular system, unspecif.	12	0,08	122718	0,042 – 0,142
Q280	AV malformation of precerebral vessels	4	0,03	368153	0007 – 0,070
Q282	AV malformation of cerebral vessels	7	0,05	210373	0,019 – 0,098
Q288	Other specif. CMs of circulatory system	10	0,07	147261	0,033 – 0,125
Q289	CM of circulatory system, unspecified	57	0,39	25835	0,293 – 0,502

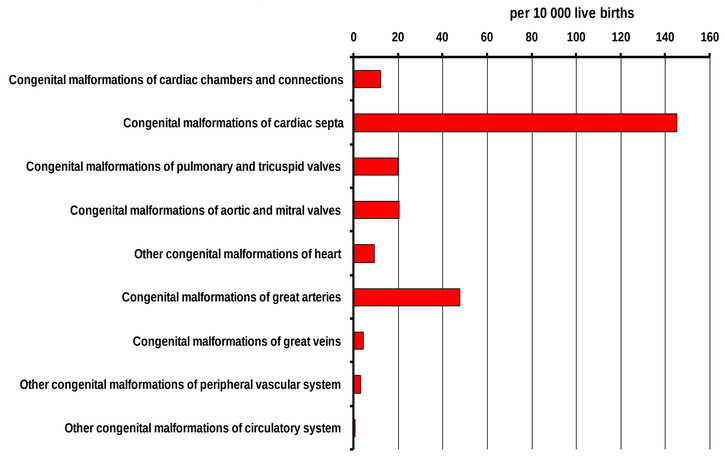
Congenital heart defects in the Czech Republic 1994 – 2008 – selected diagnosis ICD X. (Q20-Q28)

ICD-X		n	Incidence	Frequency	95% CI
Q200	Common arterial trunk	32	0,22	46019	0,149 - 0,307
Q201	Double outlet right ventricle	69	0,47	21342	0,365 - 0,593
Q203	Discordant ventriculoarterial connection	34	0,23	43312	0,160 - 0,323
Q204	Double inlet ventricle	19	0,13	77506	0,078 - 0,201
Q205	Discordant atrioventricular connection	12	0,08	122718	0,042 - 0,142
Q206	Isomerism of atrial appendages	12	0,08	122718	0,042 - 0,142
Q208	Other CM of card. chambers and connect.	67	0,45	21979	0,353 - 0,578
Q209	CM of card. chamb. and conn., unspecif.	25	0,17	58904	0,110 - 0,251
Q210	Ventricular septal defect	137	0,93	10749	0,781 - 1,100
Q211	Atrial septal defect	15	0,10	98174	0,057 - 0168
Q212	Atrioventricular septal defect	120	0,81	12272	0,676 - 0,974
Q213	Tetralogy of Fallot	26	0,18	56639	0,115 - 0,259
Q220	Pulmonary valve atresia	46	0,31	32013	0,229 - 0,417
Q221	Congenital pulmonary valve stenosis	35	0,24	42075	0,166 - 0,331
Q226	Hypoplastic right heart syndrome	23	0,16	64027	0,099 - 0,234
Q228	Other CMs of tricuspid valve	23	0,16	64027	0,099 - 0,234
Q229	CM of tricuspid valve, unspecified	11	0,07	133874	0,037 - 0,134
Q230	Congenital stenosis of aortic valve	39	0,26	37759	0,188 - 0,362
Q232	Congenital mitral stenosis	20	0,14	73631	0,083 - 0,210
Q234	Hypoplastic left heart syndrome	139	0,94	10594	0,121 - 0,267
Q238	Other CMs of aortic and mitral valves	27	0,18	54541	0,081 - 0,285
Q239	CM of aortic and mitral valves, unspecif	12	0,08	122718	0,042 - 0,142
Q240	Dextrocardia	26	0,18	56639	0,115 - 0,259
Q248	Other specified CMs of heart	38	0,26	38753	0,183 - 0,354
Q249	CM of heart, unspecified	54	0,37	27271	0,275 - 0,478
Q251	Coarctation of aorta	35	0,24	42075	0,166 - 0,331
Q252	Atresia of aorta	29	0,20	50780	0,132 - 0,283
Q254	Other congenital malformations of aorta	28	0,19	52593	0,126 - 0,275
Q255	Atresia of pulmonary artery	13	0,09	113278	0,034 - 0,143
Q258	Other CMs of great arteries	43	0,29	34247	0,211 - 0,393
Q259	CM of great arteries, unspecified	14	0,10	105186	0,052 - 0,160

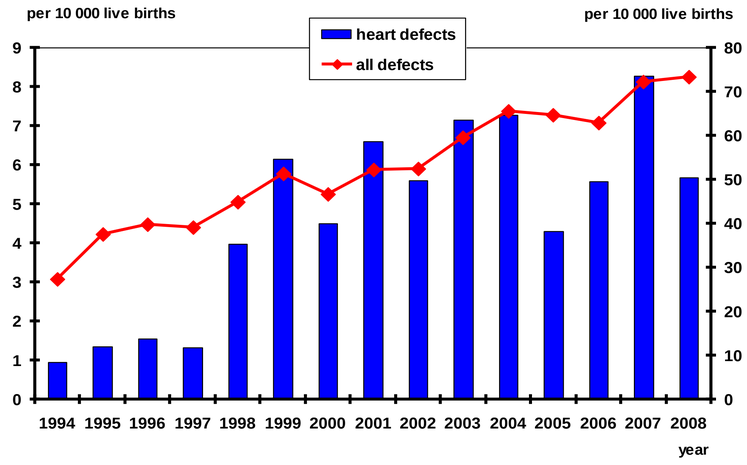
Incidence of heart defects and all birth defects in the Czech Republic, 1994 – 2008, in births



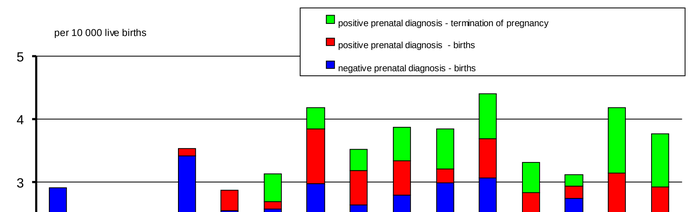
Incidence of heart defects and all birth defects in the Czech Republic , 1994 - 2008 by groups ICD X. (Q20-Q28), in births



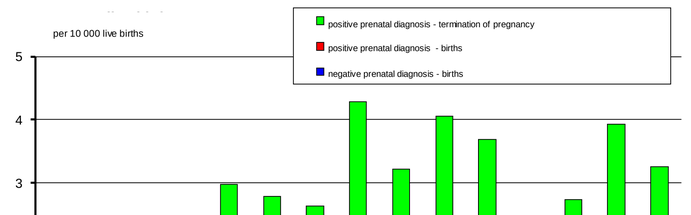
Incidence of heart defects and all birth defects in the Czech Republic 1994 – 2008 – prenatal diagnosis



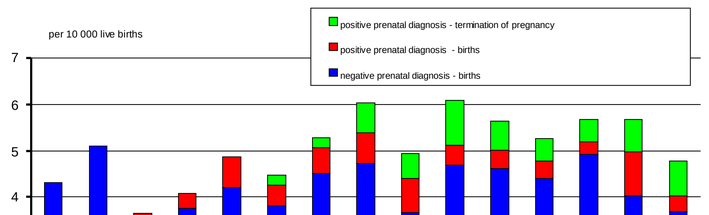
Incidence of discordant ventriculoarterial connection in the Czech Republic, 1994 – 2008



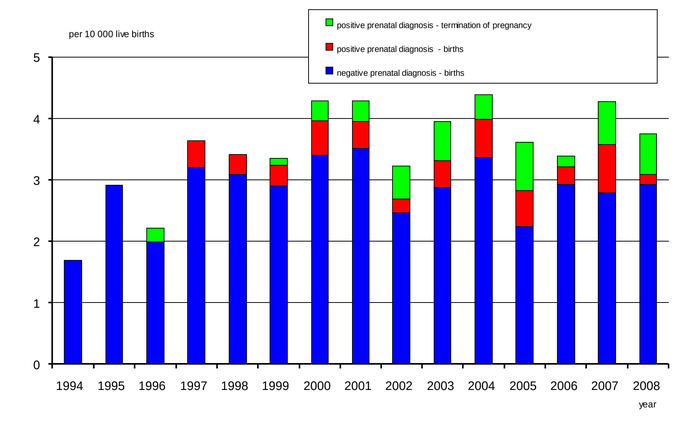
Incidence of hypoplastic left heart syndrome in the Czech Republic, 1994 – 2008



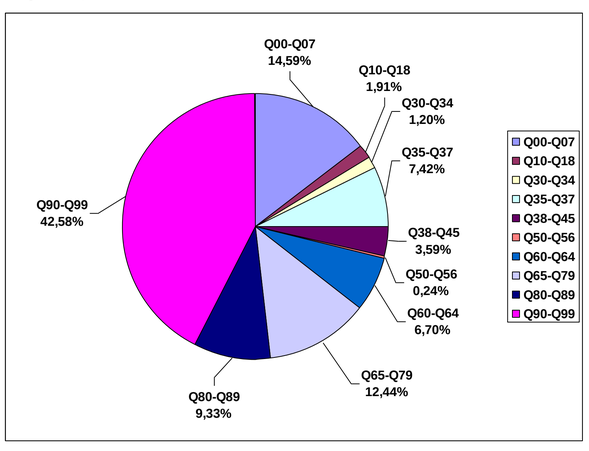
Incidence of coarctation of aorta in the Czech Republic, 1994 – 2008



Incidence of tetralogy of Fallot in the Czech Republic, 1994 – 2008



Associated malformations in prenatal diagnosis of congenital heart defects: Czech Republic, 1994 - 2008



Congenital malformations of the nervous system (Q00-Q07),
Congenital malformations of eye, ear, face and neck (Q10-Q18),
Congenital malformations of the circulatory system (Q20-Q28),
Congenital malformations of the respiratory system (Q30-Q34),
Cleft lip and cleft palate (Q35-Q37),
Other congenital malformations of the digestive system (Q38-Q45),
Congenital malformations of genital organs (Q50-Q56),
Congenital malformations of the urinary system (Q60-Q64),
Congenital malformations and deformations of the musculoskeletal system (Q65-Q79),
Other congenital malformations (Q80-Q89) and
Chromosomal abnormalities (Q90-Q99).

Groups of birth defects (death causes) - mortality in births with congenital heart defects in the Czech Republic, 1994 – 2008