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Incidence of Congenital Heart Defects in Budapest

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In $7.06 \pm 0.91‰$ of the total number of births registered in Budapest during the years 1963 to 1965, there occurred some congenital heart defects. In spite of the complex system of data collection (the data contained in the records of all Budapest institutes of pathology, cardiac surgery and paediatric cardiology as also those of the Budapest Infantile Mortality Register and the Hungarian Congenital Malformation Register were surveyed) $7.06‰$ should be regarded as the lowest possible incidence. For more than half (52.3%) of all congenital heart defects VSD, ASD and PDA were responsible, while ten anomalies formed the background of 94.6% of the registered defects. Fifty-nine per cent of these patients died before school age. The descending order of gravity was, truncus communis, transposition of the aorta and of the pulmonary artery, ostium atrioventriculare communis, Fallot's tetralogy, persistent ductus arteriosus (?), ventricular septal defect, pulmonary stenosis, coarctation of the aorta, aortic stenosis, atrial septal defect. Epidemiological data spoke for the assumption that each type represents a nosological type different in origin. Heart defects were found to constitute the most frequent and one of the gravest congenital malformations.

Values for the incidence of congenital heart defects, as reported in literature, vary over a wide range, a fact mainly due to the difficulties of examination and the diagnostic pitfalls involved [6, 16]. Let us point in this respect to (i) differences in the definition of congenital malformations; (ii) differences in the population examined (stillborn–liveborn, necropsy–clinical material, age differences); (iii) differences in diagnostic methods (anatomical, surgical, cardiological, general clinical); (iv) differences in the collection and analysis of data concerning index patients; (v) differences regarding the attitude, competence and conscientiousness of the examiners. The improvement of the technical level of epidemiological studies is illustrated by the fact that authentic examinations carried out in the 1950s showed an incidence of 3 to $4‰$, whereas those made in the sixties showed the incidence of congenital heart disease to be 6 to $8‰$ [4, 14].

The present study had the purpose to establish the incidence of congenital heart disease among the Budapest children liveborn or stillborn in the years 1963, 1964 and 1965. We have, therefore, analysed the data contained in the records of all the institutes of pathology, cardiac surgery and paediatric cardiology, further the

Budapest Infantile Mortality Register as well as the Hungarian Congenital Malformation Register. All stillborn babies as also dead infants and children were necropsied so that congenital malformations could be detected. The surviving children were 5 to 7 years old at the time of the study (second half of 1970) and thus clinically manifest congenital anomalies must have been discovered by that time. It follows that — in principle at least — the population examined by us ought to include the overwhelming majority of cases of congenital malformations.

Material and Method

(1) *Definition*. Congenital heart disease is understood to mean some gross structural abnormality of the heart or of the intrathoracic major vessels. Our classification followed the VIIIth revision of the International Classification of Diseases (ICD). The suggestions concerning heart diseases in childhood, advanced at Groningen in 1970 by the International Society of Cardiology [9], could not yet made use at the time of preparing this study. With a view to a possible differentiation between the nosological units we distinguished the following categories within the ICD groups: isolated (i.e. only the specified) heart defects; combined defects (i.e. the simultaneous existence in the heart of several anomalies); multiple anomalies (in which some heart defect is associated with some non-cardiological anomaly). On the other hand, combined heart defects occurring in just one or two cases have not been taken into account as separate groups; only the most grave types have been considered. (Our material does not include 23 cases in which a patent foramen ovale was diagnosed post mortem.)

(2) *Population*. This term covers the liveborn and stillborn babies delivered by inhabitants of Budapest in 1963, 1964 and 1965. The incidence of index patients, collected from various sources, is thus referred to the total number of births during the said years. Migration of the Budapest population was negligible in these years.

(3) *Diagnosis* made post mortem or at surgical interventions or those based on up-to-date cardiological examinations (catheterization, etc.) may be accepted as reliable [17]. Special diagnostic problems arising in certain types of congenital heart defects will be discussed further below.

(4) For tracing and recording of *index patients* we have surveyed the post-mortem reports of 18 metropolitan institutes of pathology and noted the data for all babies stillborn between January 1, 1963, and December 31, 1965, further the particulars of all babies who were born alive during this period but had died before June 30, 1970, if a congenital heart defect was revealed by the post-mortem examination. (We recorded not only the diagnosis but also the description of the heart because this procedure promised to make subsequent evaluation more uniform.) We also analysed the data contained in the register of dead Budapest infants (kept by the Pál Heim Children's Hospital). Data regarding non-fatal congenital malformations were collected from the records of four Budapest institutes of cardiac surgery. We received moreover a list from the Budapest Centre of Paediatric Cardiology containing the names of registered congenital cardiopaths. It included the results of pre-school examinations in respect of the 1963 and partly those of the 1964 age group. Only the exactly diagnosed cases of congenital heart disease were considered from this source. We also surveyed the Budapest cases recorded in the Hungarian Congenital Malformation Register. It follows from the manner of collection that a certain heterogeneity of the data regarding index patients had to be taken into account.

The data collected were entered on personal cards so that repeated registration

of the same patient was avoided. We took care to elucidate details in connexion with other anomalies. Suitably instructed medical students called on the parents of all index patients in order to verify the personal data and the diagnosis. Doubtful data were further examined and, as a result, we excluded 27 index patients from the material.

The secular trend was calculated by means of the formula

$$\chi^2_{k-1} = \sum_{i=1}^{k} \frac{\left(v_i - N_i \frac{v}{N}\right)^2}{N_i \frac{v}{N}\left(1 - \frac{v}{N}\right)}$$

where k = number of the years examined; v_i and v = the annual and the total number of congenital malformations, N_1 and N = the annual and the total number of births. The sex ratio was determined in the customary way, $\frac{\text{boys}}{\text{boys} + \text{girls}}$.

Results and Discussion*

Referred to the total number of births in Budapest during the years 1963 to 1965, the incidence of congenital heart defects amounted to $7.06 \pm 0.91‰$ (Table I). The incidence would have amounted to $7.49‰$ had the cases of patent foramen ovale been included, and to $6.98‰$ for live births. These figures are essentially in harmony with the corresponding statistical data for advanced countries. Certain distorting factors — some tending to augment the incidence value slightly, others tending to reduce it considerably — may have influenced the result so that the figure $7.06‰$ probably indicates the minimum incidence. If the increased intrauterine death rate is taken into account, the actual incidence of congenital heart defects may in our opinion amount to $8.5-9.0‰$. Since the incidence of congenital heart defects in Budapest ($7.06‰$) is presumably not essentially different from the average for the whole country, it is at every 140th delivery that the birth of an offspring with cardiac anomaly may be expected, which means that about 1100 babies are born with congenital heart defect in Hungary per year and thus heart defects constitute the largest group of congenital malformations.

The annual incidence displayed varying values in the years examined; the difference — just above the threshold of statistical significance — was due to the higher value for the year 1963, possibly a result of differences in the methods of registration [11], for instance, the effect of the cardiological screening of school-age children born in 1963.

For more than half (52.3%) of the congenital heart defects, three types were responsible, i.e. VSD, ASD and

*List of abbreviations

TC = truncus communis
TAP = transposition of aorta and pulmonary artery
FT = Fallot's tetralogy
VSD = ventricular septal defect
VSD + PS = Fallot's tetralogy without cyanosis
ASD = atrial septal defect
OAC = common atrioventricular ostium
AV = atrioventricular valvular malformation
EF = endocardial fibroelastosis
PDA = persistent ductus arteriosus
CA = coarctation of aorta
AS = aortic stenosis
PS = pulmonary stenosis
PS + ASD = Fallot's trilogy
VR = irregularities of major veins

TABLE I

Incidence, distribution and sex ratio of the various types of congenital

I. C. D. code No.	Type of defect	1963		1964	
		No.	per thousand	No.	per thousand
746.0	TC* isolated	6	0.36	2	0.11
	multiple	3	0.18	1	0.06
	total	9	0.54	3	0.17
746.1	TAP isolated	6	0.36	3	0.17
	multiple	0	0.00	0	0.00
	total	6	0.36	3	0.17
746.2	FT** isolated	3	0.18	9	0.51
	multiple	1	0.06	0	0.00
	total	4	0.24	9	0.51
746.3	VSD isolated	17	1.01	30	1.71
	multiple	11	0.66	6	0.34
	total	28	1.68	36	2.05
	VSD + PS *only* isolated	4	0.24	5	0.28
	VSD total	32	1.92	41	2.33
746.4	ASD isolated	17	1.01	13	0.74
	multiple	4	0.24	3	0.17
	total	21	1.23	16	0.91
746.5	OAC *only* multiple	5	0.30	11	0.63
746.6	AV	2	0.11	1	0.06
746.7	EF⊕	1	0.06	0	0.00
746.8	Other specified heart defects: dextrocardia	1	0.06	0	0.00
	cor biloculare	0	0.00	1	0.06
	hypoplasia of left heart	1	0.06	2	0.11
	total	2	0.11	3	0.17
746.9	Not specified heart defects	1	0.06	4	0.23
747.0	PDA^{++} isolated	14	0.84	12	0.68
	multiple	3	0.18	0	0.00
	total	17	1.02	12	0.68
747.1	CA^{+++} isolated	8	0.48	6	0.34
	multiple	1	0.06	0	0.00
	total	9	0.54	6	0.34

heart defects among infants born in Budapest in 1963, 1964 and 1965

1965		1963—1965			Secular (annual) trend	Percentage of all congenital heart defects	Sex distribution		Sex ratio
No.	per thousand	No.	per thousand	%			male	female	
1	0.05	9	0.17	64.3			4	5	0.4444
1	0.05	5	0.10	35.7	$\chi^2_2 = 6.95$		4	1	0.8000
2	0.11	14	0.27	100.0	$p < 0.01$	3.77	8	6	0.5714
6	0.34	15	0.29	93.8			9	6	0.6000
1	0.05	1	0.02	6.2	$\chi^2_2 = 1.57$		1	0	1.0000
7	0.38	16	0.31	100.0	$p > 0.05$	4.30	10	6	0.6250
7	0.38	19	0.36	86.4			12	7	0.6315
2	0.11	3	0.06	13.6	$\chi^2_2 = 1.89$		2	1	0.6666
9	0.51	22	0.42	100.0	$p > 0.05$	5.93	14	8	0.6363
26	1.48	73	1.39	75.3			42	31	0.5753
7	0.40	24	0.46	24.7	$\chi^2_2 = 0.67$		11	13	0.4583
33	1.88	97	1.85	100.0	$p > 0.05$	26.15	53	44	0.5463
1	0.06	10	0.19	100.0	$\chi^2_2 = 2.80$ $p > 0.05$	2.69	6	4	0.6000
34	1.93	107	2.04	—	$\chi^2_2 = 1.17$ $p > 0.05$	(28.84)	59	48	0.5514
9	0.51	39	0.74	81.2			26	13	0.6666
2	0.11	9	0.17	18.8	$\chi^2_2 = 4.11$		3	6	0.3333
11	0.60	48	0.91	100.0	$p > 0.05$	12.94	29	19	0.6041
4	0.22	20	0.38	100.0	$\chi^2_2 = 4.33$ $p < 0.05$	5.39	7	13	0.3500
0	0.00	3	0.06	—	—	0.81	0	3	0.0000
1	0.06	2	0.04	—	—	0.54	1	1	0.5000
1	0.05	2	0.04	—			2	0	1.0000
0	0.00	1	0.02	—			1	0	1.0000
1	0.05	4	0.08	—			2	2	0.5000
2	0.11	7	0.14	—	—	1.89	5	2	0.7142
3	0.16	8	0.15	—	—	2.16	4	4	0.5000
15	0.82	39 (28)	0.74 (0.53)				17	22	0.4358
1	0.05	6 (4)	0.12 (0.08)				2	4	0.3333
16	0.87	45 (32)	0.85 (0.61)	100.0	$\chi^2_2 = 1.13$ $p > 0.05$	12.13 (8.63)	19	26	0.4222
3	0.17	17	0.32				10	7	0.5882
3	0.17	4	0.08		$\chi^2_2 = 0.60$		2	2	0.5000
6	0.34	21	0.40	100.0	$p > 0.05$	5.66	12	9	0.5714

Table I (cont.)

I. C. D. code No.	Type of defect	1963		1964	
		No.	per thousand	No.	per thousand
747.2	AS isolated	9	0.54	6	0.34
	multiple	1	0.06	0	0.00
	total	10	0.60	6	0.34
	Other aortic defects	3	0.18	0	0.00
747.3	PS *only* isolated	8	0.48	4	0.23
	PS + ASD *only* isolated	2	0.12	1	0.06
	PS, total	10	0.60	5	0.28
747.4	VR	3	0.18	1	0.06
746–747	Heart defects total	135	8.08	121	6.89

* 4 cases of TC associated with cor biloculare.
** Fallot's pentalogy in 2 cases.
*** 3 further cases of AV were transferred to other groups.
+ 6 further cases of EF were transferred to other groups (AS three, CA two, AV one).

PDA, whereas ten types (or their combinations) were responsible for 94.6% of all congenital cardiac anomalies (Table II).

The outcome of the various types of congenital heart defects is listed in Table III. The gravity of a type is best characterized by the ratio of fatalities and survivals. The lethality rate in descending order was TC, TAP, OAC, FT, PDA (?), VSD, PS, CA, AS, ASD; 59% of these children died before school age. As a rule, the lethality rate diminishes with advancing age. In Hungary, 42% of all deaths, which occurred in 1970 in consequence of congenital anomalies, were due to heart and vascular defects. The lethality of multiple anomalies was still higher [2].

Analysis of the epidemiological parameters — details of which have been discussed elsewhere [1] — points to that the various types of heart defect are nosological units of various origin. It is therefore erroneous to study the heart defects in their entirety; the correct procedure is to determine the incidence, the epidemiological and genetic features of each nosological type separately. Examinations governed by this principle will only yield reliable values for their incidence. It should be borne in mind that VSD may disappear spontaneously, the very reason why it has the highest frequency in the newborn. ASD and mild forms of AS are generally detected late so that their registered neonatal incidence is lower than in reality. Detection of CA requires a particularly thorough pathological experience.

Examinations made along the said

1965		1964–1965			Secular (annual) trend	Percentage of all congenital heart defects	Sex distribution		Sex ratio
No.	per thousand	No.	per thousand	%			male	female	
11	0.60	26	0.49	86.7			18	8	0.6923
3	0.16	4	0.08	13.3	$\chi^2_2 = 2.86$		1	3	0.2500
14	0.77	30	0.57	100.0	$p > 0.05$	8.09	19	11	0.6333
1	0.05	4	0.08	—	—	1.08	3	1	0.7500
3	0.16	15	0.29	100.0	$\chi^2_2 = 3.34$ $p > 0.05$	4.04	7	8	0.4666
2	0.11	5	0.10	100.0	—	1.35	2	3	0.4000
5	0.27	20	0.39	—	$\chi^2_2 = 3.07$ $p > 0.05$	(5.39)	9	11	0.4500
0	0.00	4	0.08	—	—	1.08	2	2	0.5000
115	6.29	371	7.06	—	$\chi^2_2 = 4.11$ $p < 0.05$	100.00	201	170	0.5417

++ For explanation of figures in bracket, see text.
+++ In 6 cases other heart defects were also diagnosed (VSD one, VSD + AV one, VSD + PDA two, VSD + ASD one, VSD + PDA + AS one).

TABLE II

Percentage distribution and outcome of the more frequent congenital heart defec ts

Defects in descending order of frequency		Material of NADAS [15]	1906 paediatric cases of the *National Institute of Cardiology*	Material of the present study		
				363 diagnosed cases	Survivals (%)*	
					1 year old	5—7 years old
1.	VSD	20.0	20.1	26.7	56.25	45.83
	(+PS)			(29.5)	(57.14)	(45.71)
2.	ASD	10.0	13.0	13.2	68.09	57.45
3.	PDA	12.3	12.2	12.4	57.78	55.62
				(8.6)	(81.25)	(78.12)
4.	AS	5.7	12.1	8.3	62.07	55.17
5.	Fallot	14.6	8.8	6.1	54.55	31.82
6.	CA	5.0	5.6	5.8	42.86	38.12
7.	OAC	3.9	4.0	5.5	21.05	10.53
8.	PS	12.0	11.9	4.1	53.33	46.67
	(+ASD)			(5.5)	(65.00)	(60.00)
9.	TAP	4.0	1.3	4.4	6.25	6.25
10.	TC	—	—	3.9	0.00	0.00
	Total	87.5	89.0	90.4 (94.6)	41.87	50.41

* Only live births.

TABLE III

Lethality of congenital

I. C. D. code No.	Type of defect	Stillborn		Death before 28 days		Perinatal	
		No.	per cent	No.	per cent	No.	per cent
746.0	TC isolated	1	11.1	4	44.4	5	55.6
	multiple	1	20.0	2	40.0	3	60.0
	total	2	14.3	6	42.9	8	57.1
746.1	TAP isolated	0	0.0	4	26.7	4	26.7
	multiple	0	0.0	1	100.0	1	100.0
	total	0	0.0	5	31.3	5	31.3
746.2	FT isolated	0	0.0	3	15.8	3	15.8
	multiple	0	0.0	1	33.3	1	33.3
	total	0	0.0	4	18.2	4	18.2
746.3	VSD isolated	1	1.4	13	17.8	14	19.2
	multiple	0	0.0	10	41.7	10	41.7
	total	1	1 0	23	23.7	24	24.7
	VSD + PS	1	10.0	1	10.0	2	20.0
	VSD total	2	1.9	24	22.4	26	24.3
746.4	ASD isolated	0	0.0	6	15.4	6	15.4
	multiple	1	11.1	5	55.6	6	66.7
	total	1	2.1	11	22.9	12	25.0
746.5	OAC isolated	0	0.0	0	0.0	0	0.0
	multiple	1	5.0	6	30.0	7	35.0
	total	1	5.0	6	30.0	7	35.0
746.6	AV	0	0.0	1	33.3	1	33.3
746.7	EF	1	50.0	0	0.0	1	50.0
746.8	Other specified heart defects						
	dextrocardia	0	0.0	2	100.0	2	100.0
	cor biloculare	0	0.0	0	0.0	0	0.0
	hypoplasia of left heart	0	0.0	3	75.0	3	75.0
	total	0	0.0	5	71.4	5	71.4
746.9	Not specified heart defects	0	0.0	2	25.0	2	25.0
747.0	PDA isolated	0	0.0	12	30.8	12	30.8
	multiple	0	0.0	2	33.3	2	33.3
	total	0	0.0	14	31.1	14	31.1
747.1	CA isolated	0	0.0	4	33.3	4	33.3
	multiple	0	0.0	2	66.7	2	66.7
	total	0	0.0	6	40.0	6	40.0
747.2	AS isolated	0	0.0	8	30.8	8	30.8
	multiple	1	25.0	2	50.0	3	75.0
	total	1	3.3	10	33.3	11	36.7

heart defects

Death after 28 days		Death after a year		Total lethality		Survivors		Total	
No.	per cent	No.	per cent	No.	per cent	No.	per cent	No.	per cent
4	44.4	0	0.0	9	100.0	0	0.0	9	100.0
2	40.0	0	0.0	5	100.0	0	0.0	5	100.0
6	42.9	0	0.0	14	100.0	0	0.0	14	100.0
10	66.7	0	0.0	14	93.3	1	6.7	15	100.0
0	0.0	0	0.0	1	100.0	0	0.0	1	100.0
10	62.5	0	0.0	15	93.8	1	6.2	16	100.0
5	26.3	4	21.1	12	63.2	7	36.8	19	100.0
1	33.4	1	33.3	3	100.0	0	0.0	3	100.0
6	27.3	5	22.7	15	68.2	7	31.8	22	100.0
10	13.7	7	9.6	31	42.5	42	57.5	73	100.0
9	37.5	3	12.5	22	91.7	2	8.3	24	100.0
19	19.6	10	10.3	53	54.6	44	45.4	97	100.0
2	20.0	2	20.0	6	60.0	4	40.0	10	100.0
21	19.6	12	11.9	59	55.1	48	44.9	107	100.0
1	2.6	4	10.3	11	28.2	28	71.8	39	100.0
2	22.2	1	11.1	9	100.0	0	0.0	9	100.0
3	6.3	5	10.4	20	41.7	28	58.3	48	100.0
0	0.0	0	0.0	0	0.0	0	0.0	0	100.0
9	45.0	2	10.0	18	90.0	2	10.0	20	100.0
9	45.0	2	10.0	18	90.0	2	10.0	20	100.0
1	33.3	1	33.3	3	100.0	0	0.0	3	100.0
1	50.0	0	0.0	2	100.0	0	0.0	2	100.0
0	0.0	0	0.0	2	100.0	0	0.0	2	100.0
1	100.0	0	0.0	1	100.0	0	0.0	1	100.0
1	25.0	0	0.0	4	100.0	0	0.0	4	100.0
2	28.6	0	0.0	7	100.0	0	0.0	7	100.0
2	25.0	0	0.0	4	50.0	4	50.0	8	100.0
4	10.3	0	0.0	16	41.0	23	59.0	39	100.0
1	16.7	1	16.7	4	66.7	2	33.3	6	100.0
5	11.1	1	2.2	20	44.4	25	55.6	45	100.0
4	33.3	1	6.7	9	52.9	8	47.1	17	100.0
2	66.7	0	0.0	4	100.0	0	0.0	4	100.0
6	40.0	1	6.7	13	61.9	8	38.1	21	100.0
1	3.8	2	6.7	11	42.3	15	57.7	26	100.0
0	0.0	0	0.0	3	75.0	1	25.0	4	100.0
1	3.3	2	6.7	14	46.7	16	53.3	30	100.0

Table III (cont.)

I. C. D. code No.	Type of defect	Stillborn		Death before 28 days		Perinatal	
		No.	per cent	No.	per cent	No.	per cent
	Aorta, other	0	0.0	2	50.0	2	50.0
747.3	PS isolated	0	0.0	3	20.0	3	20.0
	PS + ASD	0	0.0	0	0.0	0	0.0
	PS, total	0	0.0	3	15.0	3	15.0
747.4	Transpos. ven. pulm.	0	0.0	0	0.0	0	0.0
	other	0	0.0	2	100.0	2	100.0
	total	0	0.0	2	50.0	2	50.0
746—747	Cong. heart defects, total	8	2.2	101	27.5	109	29.4

lines pointed, on the other hand, to the possibility that types regarded as separate may be identical nosologically. It is, for example, reasonable to suppose that VSD + PS and TF, and there is no doubt that the ostium primum type of ASD and OAC, are just different manifestations of anomalies originating from the same source. It is highly probable that the isolated and the multiple types of congenital defects belong to fundamentally different categories. Isolated cases usually represent separate nosological types, whereas multiple cases often represent combinations of syndromes with different aetiologies [2].

The most frequent type of heart defect is VSD in Hungary too. Its incidence of $1.85^0/_{00}$ is in harmony with the recent findings of MITCHELL et al. [14] in respect of the USA. The degree of VSD varies from Roger's disease to the cor monoventriculare biatriatum [8]. The defect closes spontaneously during the first years of extrauterine life in 25 to 50% of the cases [10, 5]. VSD is often associated with other heart defects, with infundibular PS in particular, but sometimes also with CA. Its incidence (including both these associations and FT) amounts to $2.45^0/_{00}$. VSD is moreover frequently (24.7%) associated with non-cardiac anomalies. These are probably not of the same origin as the isolated cases; they may, for instance, be partial manifestations of Down's or Patau's syndrome which originate from chromosomal aberrations. Combined anomalies are, on the other hand, rare with VSD + PS and the FT groups which are characterized by the predominance of male patients.

The incidence of ASD amounts to $0.91^0/_{00}$. Its components are different in origin [3]. Some 10 to 25% of these cases represent the so-called ostium-primum type resulting from a partial occlusion of the arterioventricular canal. The majority of the remainder belongs to the ostium secundum type. We were not in a position to examine the two types separately. It was in 18.8% of our material

Death after 28 days		Death after a year		Total lethality		Survivors		Total	
No.	per cent	No.	per cent	No.	per cent	No.	per cent	No.	per cent
1	25.0	1	25.0	4	100.0	0	0.0	4	100.0
4	26.7	1	6.7	8	53.3	7	46.7	15	100.0
0	0.0	0	0.0	0	0.0	5	100.0	5	100.0
4	20.0	1	5.0	8	40.0	12	60.0	20	100.0
1	50.0	0	0.0	1	50.0	1	50.0	2	100.0
0	0.0	0	0.0	2	100.0	0	0.0	2	100.0
1	25.0	0	0.0	3	75.0	1	25.0	4	100.0
79	21.3	31	8.6	219	59.0	152	41.0	371	100.0

that ASD appeared in combination with non-cardiac anomalies. The sex ratio shows characteristic differences in the isolated and complex forms of ASD as well as in VSD.

The incidence of PDA ($0.68^0/_{00}$) compares closely with international data. Following in this respect the earlier international usage, we classified as malformation the ductus arteriosus if it was still patent at 3 months. Since obliteration of the duct is usually complete by the 18th day of life, certain authors [13, 14] regard Botallo's duct as anomalous if it is still patent after three or even two weeks. It is likewise considered an anomaly if the diameter of the ductus arteriosus exceeds that of the pulmonary artery in the newborn. Obliteration of the duct being slower in prematures, its patency should be classified as anomalous only if still present after six months [7]. It was in 17 cases of our material that in a newborn PDA was diagnosed post mortem. Diagnosis is not infallible in such cases: if they are disregarded, the incidence drops to $0.61^0/_{00}$. A female predominance is characteristic.

We were not in the position to differentiate between the more frequent valvular form of PS and its less frequent infundibular variant. The incidence of isolated PS was $0.29^0/_{00}$, i.e. somewhat lower than the expected value. The incidence was, however, high ($0.99^0/_{00}$) if its combinations with VSD (including FT) and ASD were also taken into account.

While the incidence of AS ($0.57^0/_{00}$) was somewhat higher than the expected value, that of CA ($0.4^0/_{00}$) remained slightly below it. We could not distinguish between the various types in these groups. The lower incidence of CA reflects the well-known difficulties of clinical and post mortem diagnosis [12].

The OAC group comprised 19 cases of Down's syndrome. In addition, Down's syndrome was associated with VSD in 11, with ASD in 4, with PDA in 2 cases and with an unelucidated heart defect in one case. Having assumed the incidence of Down's

disease to be 1.7‰ of which 40% were supposed to be cases of congenital heart defect, the expected number of Down's syndrome in our material was 36; that the actual number totalled 37 shows the accuracy of our method.

TC (0.27‰) and TAP (0.31‰) showed higher incidences than expected.

Epidemiological analysis of congenital cardiopathies in Budapest yielded the following results.

(i) The minimum incidence amounted to 7.06 ± 0.91‰ which must still be regarded as high, so that heart defects constituted the most frequent and one of the gravest congenital anomalies. This anomaly may be expected to occur at every 140th delivery, and 59% of the affected children died before the school age.

(ii) The characteristic epidemiological parameters (e.g. sex ratio) justify the assumption that the various types of congenital heart defects represent nosological units different in origin so that it is advisable to examine them separately.

(iii) It is highly probable that the aetiology of isolated anomalies is different from that of multiple ones.

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