

FREQUENCY AND FORMS OF INNATE MALFORMATIONS

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The problem of innate abnormalities of various organs and systems, being of biological and social importance, is still arousing interest. Nowadays, more and more new teratogenic factors are added to their own list.

Bibliographical data of the frequency of innate malformations are differently considered and analysed in various years. Thus, in 1830—1833 their percent is 0,033% (Geoffroy Saint Hilaire); 1908 it is 1,0% (Schwalbe); 1963 — 0,074—1,98% (Langmann, Prudnikova et al.); 1956—1959—1966 — 1,64% (Czabac). In 1954 7% of all new-born babies in New York have certain abnormalities (P. G. Svetlov — 1962). In 1946 death of children with malformations is 3 times more often than death after infectious diseases (Kirchmair, USA).

There are recent data of increased number of innate disorders. Rudder (1959) reports that they are 7 times more often than the period before the Second World War. Ravina et al. (1954) shows in their study the highest percent (1,9%) of malformations compared to the previously investigated 0,61%.

Based on the aforementioned facts we decided to determine the frequency of innate malformations in the autopsy material of our Department of Patho-anatomy (Higher Institute of Medicine, Varna city) for the period 1960—1974. The frequency was studied annually, thus regarding the number of cases for each year and its variability.

All autopsy dissections were analysed divided in 2 age groups: I — 0—2 years, II — 2—87 years.

It was established that innate malformations were 8,37% of all 9330 dissected patients; their percent in children of group I is 21,16%.

Innate disorders affecting only one system of the organism were most often (63,64%), followed by those affecting several systems (36,36%).

Concerning group II it was established that there was a priority of the single abnormalities (86,44%), whereas group I indicated higher percent of multiple malformations (38,70%).

Regardless of the age the heart was most often affected organ (20,36%), followed by the stomach, intestines and liver (18,74%), vessels (16,37%) and nervous system (8,79%).

Table 1
Malformation frequency (age+systems)

System Age	Heart	Digestive	Nervous	Uro-genital	Respiratory	Muscular-Skeletal	Umbilical	Tumour	Skin	Vessel
0— 2 years	20,92	18,78	13,97	9,53	5,48	8,67	2,40	2,37	1,04	16,84
2—87 years		19,80	18,70	3,62	3,39	3,80	—	—	2,99	15,90

There was a considerable difference between the studied innate disorders of both groups (table 1). Malformations of the nervous system of children up to 2 years old were 13,97%, whereas they were only 3,61% for the second group. Abnormalities of uro-genital system were 31,8% (group II) compared to 9,53% (group I). Disorders of the muscular-skeleton system in adults (group II) were only 3,80% which was rarer than those in children up to 2 years (8,67%).

Cardiac malformations in children of group I were as follows: Morbus Rouger (23,40%), Cor trilobulare (11,06%), Fallot tetrad (7,02%). Group II showed certain differences: Fallot tetrad (21,70%), Interseptal chamber defect (17,39%), Fallot triad (13,04%).

Persistent Botali duct was most often vessel disorder in group I (8,29%), then came pulmonal artery stenosis (4,25%) and vessel transposition (4,04%). Group II had another order: Persistent Botali duct and pulmonal artery stenosis (13,04%), aorta coarctation (4,34%).

Considering the reasons for death it can be pointed that 17,79% of the investigated children were still-born. Others lived only several days after labour. 29,80% lived 1 year and another 3,67% — 2 years. 47,03% of the studied children died as a result of their innate malformations. The same reason for group II concerned only 34,20%, whereas the rest 65,80% died after diseases of other various origin.

Combination with toxoplasmosis (24 cases), cytomegalia (13 cases) and mucoviscidosis was established in some of the investigated children.

We had for another object of our study to establish an eventual gradation of lethal cases with innate malformations during the recent years. More and more women work now in various enterprises with various dangerous materials. Our investigation included exclusively the abnormalities in children up to 2 years old. Since 1969 the percent of lethal cases with innate disorders is unchanged (20,96—22,87%), but when compared to that of previous years (16,21—18,78%) it is considerably higher. There is an unexplainable exception of 26,12% (1964) and 25,00% (1968).

We considered retrospectively all etiopathogenetical factors causing abnormalities. Regretfully, only 140 cases of all 708 lethal ones (group I) showed (according to the clinical records) definite data of pregnancy peculiarities and diseases of the mothers. There was a variety of reasons for malformations: nephropathia (22 cases), toxicosis (7 cases), Rh-incompatibility (21 cases), previous still-born babies, spontaneous abortions, new-born babies with abnormalities, etc. Next to those reasons came: influenza (13 cases), rubeolla (also contacts with rubeolla patients — 5 cases), mercury intoxication (1 case), lead intoxication (1 case), petrol vapours (1 case), prolonged antibiotic application (2 cases). There was no information for greater number of cases concerning any relation between malformation and its etiology. It must be pointed that analysing only retrospective data, without additional inquiries about other diseases of the mother, her professional orientation and family genetic investigation, can not allow any conclusions.

Bibliographical data of death with innate disorders are incomparable because of the different age groups. Thus, our results (group I) show cardiac abnormalities in 20,92% and vessel malformations in 16,84% of the investigated children. Richards and McMannop report 25,35% cardio-vascular disorders in children died in their first month of life, whereas Bl. Konstantinova establishes 30,3%; however, the children under her study are up to 14 years

old. She reports 6% Morbus Rouger cases of all investigated abnormalities and 20% of only cardiovascular ones (age 0—14 years). Our data show 23,40% Morbus Rouger cases in group I and 17,39% in group II. Fallot tetrad is 7,02% in our group I, whereas Abbott (1936) reports 15% for the same age.

In conclusion we establish the innate malformations, specially in perinatal and early childish periods, are an often finding in our autopsy material. Our data require a further and detailed analysis concerning reasons of abnormal foetal development, influence of house and professional factors, pregnancy and its pathology, etc.

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ЧАСТОТА И ФОРМЫ ВРОЖДЕННЫХ УРОДСТВ

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РЕЗЮМЕ

Настоящее исследование охватывает период с 1960 по 1974 г. г. Из вскрытых 9330 случаев в возрасте от 0 до 87 лет врожденные уродства составляют 8,37%. Их число значительно больше в группе от 0 до 2-летнего возраста (21,16%). Первое место по частоте занимают уродства сердца — 20,36%; уродства желудочно-кишечного тракта составляют 18,7%, сосудов — 16,37%, нервной системы — 8,79%.

Наблюдались значительные различия в частоте аномалий нервной системы и органов выделения. Так у детей в возрасте до 2 лет устанавливаются уродства в 13,97% всех вскрытых, а у пожилых — в 3,61%. Аномалии органов выделения у детей в возрасте до 2 лет составляют 9,53%, а в возрасте старше 2 лет их 31,80%.

Аномалии нескольких систем были установлены в 36,36%. В последние годы увеличение случаев с врожденными аномалиями не наблюдается.