

STATISTICS OF CONGENITAL CARDIAC DISEASE.

*(400 Cases Analyzed.)\**

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The subject of congenital cardiac disease is one that lends itself well to statistical study, for the conditions, being often complex and of recognized rarity, are usually reported in much detail. Moreover, the cases are so infrequent in any one person's experience that some such method as this, of making use of the available literature, must be adopted in order to arrive at any generalizations.

For another purpose, I have had occasion to make a detailed statistical study of some four hundred and twelve cardiac defects. A few of these are drawn from personal experience, the remainder from the literature. Only well-authenticated cases with post-mortem report attached have been included. The only exception to this statement is formed by three cases included in the series of patent ductus arteriosus diagnosed by characteristic physical signs and by the X-Rays and not confirmed by post-mortem. The results of the analysis of four hundred of these cases are shown in the accompanying chart. This chart is presented here merely as a demonstration of the manner in which these defects were studied, and without any intention of entering at length into the figures. It represents a chart which was originally printed for the analysis of the individual defect, and is here modified in a few particulars to admit of the presentation of the total results obtained.

The chart presents four main divisions. The First Division includes the Classification of the defect, Number of cases analyzed, Age, and Sex. In the classification a simple anatomical order has been followed, based also on the

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principles of the development of the heart, so far as these are known. Thus, defects of the cardiac and aortic septa are followed by transposition of the arterial trunks, due (according to Rokitansky) to a deviation of the aortic septum; and this again by pulmonary and aortic stenosis or atresia (some cases of which are probably likewise due to a deviation of the aortic septum). The cases of pulmonary and aortic stenosis or atresia are sub-classified (following Rauchfuss) according to the presence or absence of defects of the interauricular and interventricular septa, and this affords a clinical grouping of much value. In coarctation of the aorta the distinction drawn by Bonnet\* is observed between the infantile form, a simple persistence of the isthmus aortæ, and the typical "adult type" of coarctation in which the aorta is obstructed or even obliterated by a sharp constriction at or above the insertion of the ductus.

Among the defects enumerated in this classification those of clinical importance are:

Defects of the interauricular septum, 28 cases; defects of the interventricular septum, 40 cases; complete defects of the cardiac septa (biloculate heart, etc.), 12 cases; defects of the aortic septum, 14 cases; transposition of the arterial trunks, 44 cases; pulmonary stenosis, 75 cases; pulmonary atresia, 23 cases; tricuspid stenosis, 2 cases; tricuspid atresia, 9 cases; patent ductus arteriosus, 23 cases; coarctation of the aorta, 33 cases; hypoplasia of the aorta, 2 cases. Under "Age" are three columns in which the maximum, minimum, and mean ages of the cases in each group are calculated.

The Second Division of the chart includes those Post-mortem Findings of especial importance in cardiac defects. Here are noted the condition of the fetal passages, whether closed or patent, the presence of hypoplasia or dilatation of the pulmonary artery or the aorta, the existence of a collateral circulation (important in coarctation of the aorta), the incidence of arterial disease, of acute endocarditis, and of

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\* Bonnet. *Revue de Médecine*, 1903.

chronic valvular disease, the presence of hypertrophy and dilatation of the different chambers of the heart, and lastly the existence of associated anomalies in the heart, vessels, or elsewhere.

The Third Division notes points of clinical interest, such as the presence of conditions having an etiological bearing on the family history, and in the personal history the incidence of rheumatism, pulmonary tuberculosis, or congenital syphilis, and the proportion of cases recovering from the acute infectious fevers (which cyanotic patients are said to pass through well). Under special symptoms are columns for cyanosis in its different degrees, clubbing of the fingers, dyspnea, dyspneic attacks and delayed development. Physical signs may be vascular or cardiac, and among the latter the occurrence of visible pulsation, precordial bulging, thrill, increased dulness, accentuation of the heart sounds, and the existence of murmurs, presystolic, systolic, diastolic, continuous, double (*i.e.*, systolic and diastolic in rhythm), or indefinitely stated, are noted. Finally, under causes of death we find the defect itself proving fatal suddenly or by failing compensation, or a termination by broncho-pneumonia, cerebral complications or the acute infectious fevers.

The Fourth Division of the chart, that of Relative Frequency, is of the greatest importance. Cardiac anomalies are so often complicated that the number of times a given defect occurs alone or as the primary condition by no means represents its total incidence in the four hundred cases. In this division there are, therefore, three columns. In the first of these stands "the number of cases classified as the primary lesion," the figures of which are identical with those at the beginning of the chart showing "the number of cases analyzed" in each group. The sum of the figures in this column is the four hundred cases analyzed. In the next column stands the number of cases in each group in which the defect occurs complicating other conditions, and this with the number of cases classified as the primary lesion

gives the total incidence of the defect, which is thus shown in the last column of the chart.

The result of this analysis brings out some remarkable facts, several of which are at variance with accepted ideas. The following points are of especial interest:

1. The frequency of defects of the interventricular septum. — While relatively rare alone (thirty-two defects at the base among the four hundred), in combination with other conditions this is seen to be the most common of all cardiac anomalies (one hundred and forty-nine among the four hundred cases); next in frequency comes patent foramen ovale, under which are included only cases of true patency, not simply a valvular or slit-like condition, with one hundred and thirty-four cases, and then patent ductus arteriosus with one hundred and six. The frequency of transposition of the arterial trunks (forty-six cases) and of pulmonary stenosis with defect of the interventricular septum (seventy-three cases) is noteworthy, while pulmonary stenosis with closed interventricular septum is relatively infrequent (seventeen cases).

2. The duration of life is seen to be relatively long in uncomplicated defects of the interauricular septum, patent ductus arteriosus, coarctation of the aorta, and pulmonary stenosis with closed interventricular septum. In pulmonary stenosis with defect of this septum the duration of life is seen to be much shorter.

3. Patency of the ductus arteriosus is seen to be rare in pulmonary stenosis, though very frequent in pulmonary atresia.

4. The right chambers chiefly are hypertrophied and dilated in defects of the interauricular septum, transposition of the arterial trunks, pulmonary stenosis and atresia. Both chambers, but chiefly the right, are enlarged in defects of the interventricular septum and patent ductus arteriosus, the left ventricle chiefly in coarctation of the aorta.

5. Acute endocarditis is seen to be relatively common in



defects of the interventricular septum at the base and in pulmonary stenosis.

6. Cyanosis was absent in most of the defects of the interauricular septum and was not "marked" in any of these cases. A moderate degree of cyanosis was fairly common in defects of the interventricular septum, a marked degree in only three cases. Marked cyanosis was seen chiefly in transposition of the arterial trunks, pulmonary stenosis with defect of the interventricular septum, pulmonary and tricuspid atresia. Cyanosis was usually slight or absent in patent ductus arteriosus and in coarctation of the aorta of the adult type. In six cases of defect of the interauricular and in four of defect of the interventricular septum, the cyanosis was "terminal," appearing only in the last few weeks of life.

7. A thrill was frequent in "pure" defects of the interventricular septum at the base, and in pulmonary stenosis with closed interventricular septum, or with defect of the interventricular septum and patent foramen ovale. A thrill was relatively rare in pulmonary stenosis with defect of the interventricular septum and closed foramen ovale.

8. In the great majority of cardiac defects the murmur, when present, was systolic in rhythm.

9. In some cases of pulmonary stenosis the pulmonary second sound was accentuated.

These are not all the conclusions to be drawn from a study of this analytical table; they are sufficient, however, to show the value of a careful and detailed tabulation of the data afforded by different observers in arriving at general deductions, such as could not legitimately be drawn from the facts in the experience of any single worker.