The Quantitative Anatomy of Cyanotic Tetralogy of Fallot

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In previous work, a method was evolved making possible the prediction of the values of 21 modalities of the heart at autopsy of a normal child of any age, weight, and height up to 15 years of age. These modalities include weight of heart and of parietal ventricular walls, thickness of ventricular walls, internal dimensions of chambers, and sizes of orifices. From the internal dimensions of the ventricles, volume indices of the chambers were calculated. This work made possible the judgment as to whether a value of any modality in a given heart was normal, probably normal, abnormal, or probably abnormal. The present work extends these methods, and applies them to the study of a group of hearts in a single entity—cyanotic tetralogy of Fallot.

Materials and Methods

Measurements of all modalities except those of weight of the parietal walls were made on 15 hearts the seat of un-operated tetralogy of Fallot with clinical cyanosis at the time of autopsy, 15 hearts the seat of tetralogy of Fallot with pulmonary atresia, 17 hearts of cyanotic tetralogy of Fallot with total repair, the patient dying within 48 hours of the procedure, and 17 hearts of cyanotic tetralogy of Fallot with old aortico-pulmonary anastomosis and total repair from patients dying within 48 hours after repair. These measurements were then compared with those expected in normal cases of the corresponding age, weight, and height. Judgments were then made as to whether each measurement fell into the normal, probably normal, abnormal, or probably abnormal range. Bar graphs were then made of the proportions of the total number of cases in each of these four categories falling into the normal, probably normal, abnormal, or probably abnormal class. The expression “there is a tendency toward” was used where there was a gradual progression toward or sudden onset of a high point in any one area.

To supplement these findings in cyanotic tetralogy of Fallot without operative procedures, the observed values of each of the 17 above modalities in this entity were compared with the expected normal values of each modality for the corresponding age with use of a 5 per cent level of confidence. This made possible judgments of (1) the state at birth and after birth, (2) the progression of lesions with advancing age, and (3) the possible tendency of certain values, although in the normal range, to fall consistently above or below the mean. Where the latter occurred, the expression “there is a slight tendency toward” was used.

Findings

In cyanotic tetralogy of Fallot without operative procedures (table 1) the heart has a tendency to be normal in weight at birth, but to be increased in weight after birth. The right ventricle has a tendency to be smaller than normal in size in some cases and larger in others. Its wall has a tendency to be normal in thickness at birth but increased in thickness thereafter. The tricuspid orifice has a slight tendency to be smaller than normal at birth and thereafter. The circumferences of the infundibulum and of the pulmonic orifice have a tendency to be small at birth, and with advancing age there is relatively greater stenosis. The left ventricle has a tendency to be smaller than normal, with a wall of normal thickness. The mitral orifice has a slight
Table 1
Unoperated Cyanotic Tetralogy of Fallot; Judgments of Tendencies in Each Modality as Gleaned from Two Types of Graphs

<table>
<thead>
<tr>
<th>Modalities</th>
<th>Bar graphs</th>
<th>Comparison of predicted and observed values with advancing age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weight of heart</td>
<td>Tendency toward increase (fig. 1)</td>
<td>Normal at birth, tendency toward increase after birth (fig. 2)</td>
</tr>
<tr>
<td>Size of right ventricle</td>
<td>Tendency toward smallness in some cases and largeness in others (fig. 3)</td>
<td>—</td>
</tr>
<tr>
<td>Thickness of wall of right ventricle</td>
<td>Tendency toward increase</td>
<td>Normal at birth, tendency to increase after birth (fig. 4)</td>
</tr>
<tr>
<td>Tricuspid orifice</td>
<td>Tendency toward normality (fig. 5)</td>
<td>Slight tendency toward smallness (fig. 6)</td>
</tr>
<tr>
<td>Circumference of infundibulum</td>
<td>Clear tendency toward smallness (fig. 5)</td>
<td>Small at birth and tendency toward greater stenosis with advancing age (fig. 7)</td>
</tr>
<tr>
<td>Pulmonic orifice</td>
<td>Clear tendency toward smallness (fig. 5)</td>
<td>Small at birth and tendency toward greater stenosis with advancing age (fig. 8)</td>
</tr>
<tr>
<td>Size of left ventricle</td>
<td>Tendency toward smallness (fig. 9)</td>
<td>—</td>
</tr>
<tr>
<td>Thickness of wall of left ventricle</td>
<td>Tendency toward normality (fig. 10)</td>
<td>Tendency to normal</td>
</tr>
<tr>
<td>Mitral orifice</td>
<td>Tendency toward normality (fig. 5)</td>
<td>Slight tendency toward smallness (fig. 11)</td>
</tr>
<tr>
<td>Aortic orifice</td>
<td>Tendency toward increase (fig. 5)</td>
<td>Normal at birth, tendency toward increase thereafter (fig. 12)</td>
</tr>
</tbody>
</table>

The tendency to be smaller than normal at birth and thereafter, while the aortic orifice has a tendency to be normal at birth but enlarged thereafter.

In tetralogy of Fallot with pulmonary atresia, the right ventricle has a tendency to be thicker, the mitral valve smaller (fig. 13), and the aortic valve larger than in simple tetralogy (fig. 13). Since the tricuspid orifice was not compared in all age groups in this type, no complete statement can be made about this orifice. The bar graph reveals a tendency toward normality (fig. 13).

After total repair, 24 to 48 hours after surgery, the heart has a tendency to be increased in weight as compared to unoperated tetralogy (fig. 14). There is a greater tendency for the right ventricle to be normal in size, even though still small (fig. 3), and its wall has a tendency to be increased in thickness, as compared to unoperated tetralogy. The left ventricle now has a tendency to be normal in size (fig. 9) and its wall thinner than normal (fig. 15). The tricuspid orifice is smaller in size, and the aortic orifice more normal in size, as compared to unoperated tetralogy (fig. 16). There is a tendency for the circumference of the infundibulum, although larger than before repair, to remain smaller than normal. The pulmonic orifice shows little change from unoperated cases (fig. 16).

After total repair following an old Potts anastomosis, the heart is still more increased in weight, as compared to cases with total repair alone. The left ventricle has a tendency to be enlarged (fig. 9), with a thickened wall.
Proportion of cases in simple tetralogy in which the heart weight fell into the normal (N), probably normal (PN— in the lesser range and PN+ in the greater range), probably abnormal (PA— in the lesser range and PN+ in the greater range), and abnormal (A— in the lesser range and A+ in the greater range).

(fig. 17). There is a tendency for the mitral orifice to be enlarged, and the aortic orifice to be still more enlarged than in unoperated cases. The circumference of the infundibulum and pulmonic orifices are about the same as in total repair without Potts anastomosis. The size of the right ventricle now has a tendency to be increased (fig. 3), with a still more

Values of the heart weight plotted against age in simple tetralogy of Fallot.

Values of the thickness of the right ventricle at the tricuspid area (RV-T) in simple tetralogy plotted against age.
thickened wall than in total repair without aorticopulmonary anastomosis.

Discussion

The quantitative data corroborate the known physiologic data. In tetralogy of Fallot with or without pulmonary atresia, we are dealing with pressure hypertrophy of the right ventricle and volume atrophy (or hypotrophy) of the left ventricle, with the corresponding smallness of the mitral orifice. The reason for smallness of the right ventricle in some cases is unknown. The enlargement of the right ventricle in some cases may be attributed to failure. To these quantitative data may be added the qualitative data of increased thickness of the wall of the right atrium with normal-sized chamber, and de-
Cyanotic Tetralogy of Fallot

Figure 9
Proportion of cases in which the volume index of the left ventricle \( (V_{LV}) \) fell into the range of the normal, probably normal, abnormal, and probably abnormal in simple tetralogy (medical cases), tetralogy with pulmonary atresia, tetralogy with total repair (operated cases), and tetralogy with previous Potts procedure and total repair.

Figure 10
Proportion of cases of simple tetralogy of Fallot in which the values of the maximum thickness of the left ventricle (LV-M), and the thickness at the apex (LV-A) fell into the range of the normal, probably normal, abnormal, and probably abnormal. The important measurement is the maximum thickness. The thickness at the apex is relatively unimportant. See reference 2.

Figure 11
Values of the circumference of the mitral orifice plotted against age in simple tetralogy of Fallot.

Figure 12
Values of the circumference of the aortic orifice plotted against age in simple tetralogy of Fallot.

Increased size of the left atrium with thinning of the wall. The total effect is an increase in weight of the heart after birth. The enlargement of the aortic orifice after birth is related to volume. The finding of a normal-sized aortic orifice at birth may be questioned, since we have found great variation of measurements at birth.¹

The tendency toward smallness of the tricuspid orifice in tetralogy of Fallot is probably associated with the overriding and hence dextroposition of the aorta with the impingement of the latter on the tricuspid orifice. This, however, is not a frank physiologic stenosis although true anatomic stenosis of this orifice is common in tetralogy. The

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tendency for the infundibular circumference and the pulmonic orifice to remain the same size despite the growth of the right ventricle indicates greater functional obstruction with advancing age. The greater enlargement of the aorta in tetralogy of Fallot with pulmonary atresia as compared to that without pulmonary atresia is related to volume.

After total repair, the increase in weight of the heart 24 to 48 hours after surgery with corresponding increase in thickness of the right ventricle is possibly related to surgical handling and imbibition of water into the myocardium. It is to be noted that the left ventricle does not show this. Instead, this chamber is increased in size with a thin wall.

This raises the question of left ventricular failure in these cases due to sudden increase in pulmonary flow, or perhaps due to obstruction of the aortic orifice, both related to surgery. It is interesting that despite the infundibulectomy and some enlargement of the
tricuspid orifice still smaller than before operation. One might anticipate some untoward effects of this change in an occasional case in the future. The change of the aortic orifice to a normal size may be a desired effect, but some aortic stenosis of importance might be produced in some cases. The changes in both of these orifices may be related to the technic involved in closure of the ventricular septal defect.

The increase in total weight of the heart in cases with total repair following old aortico-pulmonary anastomosis over that in total repair alone is due to the volume hypertrophy of the left ventricle produced by the previous Potts procedure. Associated is the enlargement of the mitral orifice. To this may be added the qualitative data of enlargement of the left atrium with or without thickness of the wall. Interesting is the relative lack of shrinkage of the tricuspid and aortic orifices after surgery in this group in contrast to that found in total repair without previous aortico-pulmonary anastomosis. The greater tendency toward enlargement of the aortic orifice may be related to the anastomosis. This enlargement remains, despite the operative closure of the ventricular septal defect. This raises the question of technical differences in the repair of the ventricular septal defect with or without previous shunting procedure. The greater muscle mass of the right ventricle in this group as compared to the operative procedure without a previous Potts procedure is not understood.

It must be pointed out that our methods of measurement of internal dimensions of the right ventricle in the normal heart and in tetralogy, and the volume indices obtained thereby, are relatively crude. Furthermore, there are differences in shape between the normal right ventricle and that in tetralogy, and perhaps they are not comparable by our present methods. We have recently succeeded in fixing the heart in the end-diastolic pressure of each individual chamber.4 This will make possible more direct measurements of volume at autopsy.
Summary

A quantitative analysis was made of hearts with tetralogy of Fallot without operative procedures, with pulmonary atresia, with total repair, and with old Potts anastomosis and total repair. Useful data thus obtained were as follows:

In simple tetralogy the left ventricle has a tendency to volume atrophy. The tricuspid orifice has a tendency to be smaller than normal.

In tetralogy with pulmonary atresia, the aortic orifice has a tendency to be larger, and the mitral orifice smaller than in ordinary tetralogy.

In tetralogy after total repair, the heart is increased in weight 24 to 48 hours later probably related to manipulation of the right ventricle. The tricuspid and aortic orifices have a tendency to be smaller than previously, probably related to the technic of closure of the ventricular septal defect. This is not the case where an aorticopulmonary anastomosis has been previously done before total repair.

References


Auricular Fibrillation

The development of knowledge about auricular fibrillation illustrates the close interdependence of clinical medicine and experimental work. Auricular fibrillation, long familiar to physiologists was first recognized in man by Cushing and Edmunds in a case under their care in 1901 and published in 1906, the fibrillation being ascribed to vagal inhibition; but their diagnosis of fibrillation was received with doubt. In 1902 Mackenzie had recognized the complete irregularity of the pulse now known to be characteristic of auricular fibrillation, and on the ground of the absence of the auricular wave in the jugular tracings argued that the auricles had ceased to contract and were paralyzed. In 1907 he abandoned the idea of paralysis, as in some cases at any rate the muscular fibres of the auricles were found by Keith to be hypertrophied, and argued that the auricular and ventricular contractions occurred simultaneously as the result of irritability of the auriculo-ventricular node, and called this "nodal rhythm." In 1910 Rothberger and Winterberg, and Lewis, by means of the electrocardiograph, proved that the auricular condition was one of auricular fibrillation.—Sir Humphry Davy Rolleston. The Harveian Oration. Great Britain, Cambridge University Press, 1928, p. 108.
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