PATHOPHYSIOLOGY AND NATURAL HISTORY
CONGENITAL HEART DISEASE

Is the aorta truly dextroposed in tetralogy of Fallot?
A two-dimensional echocardiographic answer

KARL ISAAZ, M.D., JEAN L. CLOEZ, M.D., FRANCOIS MARCON, M.D., ANNE M. WORMS, M.D.,
AND CLAUDE PERNOT, M.D.

ABSTRACT  The embryogenesis of tetralogy of Fallot is still much debated. In particular, the
dextroposition of the aorta is not considered by all pathologists as a genuine abnormality in this
congenital heart disease but rather as a false impression due to an exaggeration of the normal overriding
caused by dilatation of the aorta secondary to abnormal hemodynamics. We used two-dimensional
echocardiography to examine the spatial position of the aortic root in 22 patients with tetralogy of Fallot
(aged 5 days to 24 years, mean 6.4 years) and in 23 normal subjects (aged 1 month to 27 years, mean
7.6 years). Using the parasternal short-axis view, we determined the percent rightward displacement of
the aortic root in relation to the plane of the atrial septum, and the relationship between the aortic cusps
and the atrial septum. We measured the value of the angle 0, which was defined as the angle between
the plane of the atrial septum and the plane of the left coronary–noncoronary commissure and leaflet
appositional plane. In the control group, the aortic root was displaced to the right by only 23.6 ± 7.6%;
the atrial septum crossed the posterior aspect of the aortic root at the middle (n = 19) or at the right half
of the posterior cusp (n = 4), and the angle 0 had a value of 43.3 ± 8.8 degrees. In the 22 patients with
tetralogy, the percent rightward shift of the aortic root was augmented to 55.5 ± 9% (p < .001) and the
atrial septum was related to the posterior commissure in 14 patients, to the left coronary cusp in two
patients, and to the left fourth of the posterior cusp in six patients; the angle 0 had a value of 9.2 ± 11.2
degrees (p < .001). In the two groups, the position of the commissure between the right coronary and
left coronary aortic cusps was similar in relation to a line passed forward from the atrial septum. We
conclude that the aorta is truly dextroposed in tetralogy of Fallot, with a rightward shift due to a
clockwise rotation of the aortic root (looking downstream), and that the axis of this rotation is the
anterior aortic commissure. Our echocardiographic findings, which confirm conclusions of previous
investigators based on pathologic anatomy, support the embryologic hypothesis of the lack of conal
rotation in tetralogy of Fallot and permit a quantitation of the effects of this morphogenetic mechanism.

IN THE PAST, the terms “dextroposition” and “over-
riding” have been used interchangeably. However,
overriding of the aorta should not be confused with
dextroposition. Overriding of the aorta over the right
ventricle occurs in the normal heart because of the
sigmoid interventricular septum1 as well as in hearts
with isolated infundibular perimembranous ventricular
septal defect2–3 and can be considered a normal anat-
omic feature.4 The term dextroposition indicates a
pathologic condition with a rightward displacement of
the aortic root.4,5 There has been considerable debate
concerning whether a true dextroposition of the aorta
exists in tetralogy of Fallot. Some investigators6,7 have
suggested that in tetralogy there is only an exaggera-
tion of the normal overriding aorta caused by dilatation
of this vessel secondary to increased aortic flow and
simulating a dextroposition. Other authors4,5,8–11 have
suggested that the condition is typified by a true dex-
troposition of the aorta with a rightward displacement
and a clockwise rotation of the aortic root, particularly
in cases with severe infundibular obstruction.8

Two-dimensional echocardiography has been used
to show the overriding aorta over the right ventricle in
patients with tetralogy of Fallot,12–15 but no criteria
have been proposed to differentiate a simple exagger-
ation of the normal overriding from a true dextropos-
ition.

In view of this uncertainty regarding one important
anatomic feature of tetralogy of Fallot, we used two-
dimensional echocardiography to examine 22 patients
with tetralogy of Fallot, recording geometric measure-
ments, and compared our results with similar measurements recorded in a control group of 23 normal subjects. The purpose of this study was to answer the questions: is the aorta truly dextroposed in tetralogy of Fallot? If this is the case, what is the mechanism of the dextroposition? Our objective was to provide echocardiographic arguments for or against one of the proposed embryologic hypothesis, the lack of conal rotation.

Materials and methods

Clinical population. We studied 49 patients prospectively by two-dimensional echocardiography, using the parasternal short-axis plane at aortic valve level. Complete examination of the aortic root with the three aortic cusps and atrial septum was possible in 45 (92%). Four patients were excluded because of incomplete results. Twenty-two patients had tetralogy of Fallot and ranged in age from 5 days to 24 years (mean 6.4 years); there were 13 male and nine female patients. Sixteen (72%) patients had undergone previous surgery, 11 corrective and five palliative operations.

The results were compared with those recorded in 23 patients without heart disease, aged from 1 month to 27 years (mean 7.6 years), including 13 male and 10 female patients. Examinations were performed with Advanced Technology Laboratories equipment and 3.0 and 5.0 MHz frequency transducers.

Imaging technique. Patients were examined while in a supine position with the transducer placed at the left sternal border or at the subclavicular region. The transducer was manually rotated to obtain a parasternal short-axis view of the aortic valve level and slight adjustments in transducer angulation were required to provide simultaneously a good visualization of the aortic root with the three aortic cusps, the atrial septum, and the pulmonary valve. Still frames were converted to hard copy.

Geometric measurements and calculations. Figures 1 and 2 show schematically the measurements and calculations we recorded in each patient. A circle circumscribing the inner side of the aortic root was constructed, as shown in figure 1, and the percent of its rightward displacement (d/a) in relation to the line CD passed forward from the aortic septum was recorded (figure 2). Because of the variable curvature of the middle part of the atrial septum, we took only the anterior and posterior insertions of this septal structure into consideration to construct the line CD. We completed the analysis of the relationships of the aortic root with the adjacent structures (atrial septum and pulmonary valve) by determining: (1) the point of intersection (r) between the line CD and the rim of the aortic root, and the aortic cusp related to this point, (2) the point of intersection (o) between the pulmonary valve and the anterior aspect of the aortic root, and the aortic cusp related to this point. To assess the mechanism of the hypothetical dextroposition, we constructed the following lines (figure 2): (1) through the commissure between the anterior aortic coronary and the noncoronary cusps (XY), (2) between the anterior aortic commissure on the inner border of the aortic root and the posterior insertion of the atrial septum (ZD), (3) between the anterior aortic commissure on the inner border of the aortic root, and the center of the circle circumscribing the inner edge of the aortic root (ZW).

These various lines defined the following angles: (1) angle θ, subtended by the lines CD and XY; this angle was conventionally considered as positive when the line XY was at the left of the line CD and negative when it was at the right; (2) angle φ, subtended by CD and ZD; (3) angle ρ defined by CD and ZW. Furthermore, we assessed the degree of left inclination of the atrial septum by measurement of the angle ε, which was defined by the line CD and the line AB, which was drawn through the posterior wall of the atria along the first 10 mm on each side of the atrial septum (figure 1). The measurement of the angle ε was necessary to interpret the results concerning the other angles (θ, φ, and ρ), which had the line CD as a common side.

Data analysis. All measurements and calculations were done by one examiner and repeated by another observer who was blinded to the findings of the first observer and whose determinations never differed from the first observer’s by more than 5%.

Statistical analysis. The data used for each of the comparisons between the two groups were subsequently submitted to statistical analysis with Student’s t test and a confidence level of 0.5%.

Results

The recorded measurements and calculations are indicated in table 1 for the patients with tetralogy of Fallot and in table 2 for the subjects in the con-

FIGURE 1. Schematic representation of the short-axis plane at aortic valve level showing the construction of the lines AB through the posterior wall of atria, CD through the anterior and posterior (D) points of insertion (Δ) of the atrial septum, and the angle ε subtended by these two lines. A circle circumscribing the inner rim of the aortic root is constructed and used for calculating the percent rightward displacement of the aorta in relation to the line CD. Point o is the intersection between the pulmonary valve (PV) and the anterior aspect of the aortic root; z represents the right coronary–left coronary commissure (the septal commissure) of the aortic valve. This diagram represents a hypothetical case where o is facing z; r is the point of intersection between the line CD and the inner rim of the aortic root. L = left coronary cusp; LA = left atrium; N = noncoronary cusp (or posterior cusp); PV = pulmonary valve; R = right coronary cusp; RA = right atrium; TV = tricuspid valve; (●) = center of the circle circumscribing the inner rim of the aortic root.
control group. The statistical comparisons are shown in Table 3.

These measurements showed that the angle \( c \) of the interatrial septum was identical in patients with tetralogy of Fallot and normal subjects (87 vs 86 degrees, NS). This finding permitted us to take the line CD of atrial septum as a reference for other measurements. In patients with tetralogy, the percent rightward displacement (d/a) of the aortic root in relation to the plane (CD) of the atrial septum averaged 55.5% (range 44% to 80%), which was significantly greater than the average value for the normal subjects 23.6% (range 7% to 39%; \( p < .001 \)). In patients with tetralogy, the atrial septum crossed the posterior aspect of the aortic root (point \( r \)) at the level of the left fourth of the noncoronary cusp in six of the 22 patients, at the right third of the left coronary cusp in two patients, and at the commissure between these two cusps in 14 patients. In the control group, point \( r \) was located at the middle of the noncoronary cusp in 19 patients and at the right half of this cusp in four patients. The value of the angle \( \theta \) was significantly reduced in patients with tetralogy (mean +9.2 degrees, range -11 to +30 degrees) when compared with that in normal subjects (mean +43.3 degrees, range +30 to +60 degrees) (\( p < .001 \)). There was no significant difference in the two groups between the average values of the angle \( \phi \) (mean 13.2 degrees, range 9 to 16 degrees in patients with tetralogy; mean 13 degrees, range 8 to 17 degrees in normal subjects). In normal subjects, the pulmonary valve crossed the anterior aspect of the aortic root at the level of the anterior aortic commissure in six patients (27%) and at the left third of the right coronary cusp in 16 patients (75%); in one patient, point \( o \) could not be located because the pulmonary valve was not seen. In patients with tetralogy, the position of point \( o \) could be determined in 12 of 22 patients; in four of these 12 patients (33%), point \( o \) was located at the anterior aortic commissure and in eight patients at the left third of the right aortic cusp (67%). We found a statistically significant difference between the value of the angle \( \rho \) in the two groups (\( p < .001 \)); in patients with tetralogy.

![Diagram](http://circ.ahajournals.org/)

**Figure 2.** Diagrammatic representations of the different echocardiographic measurements. Top left, Diagram showing how the rightward displacement of the aorta is assessed; \( d \) is that portion of the diameter of the aortic root that lies to the right of the plane of the atrial septum (CD), and \( a \) is the diameter of the aortic anulus. The rightward shift of aortic root is expressed as the ratio \( d/a \). Top right, Diagram defining the angle \( \theta \), which is considered by convention as less than 0 degrees when the line XY is at right of the line CD, greater than 0 degrees when it is at left, and 0 degrees when the lines CD and XY are superimposed. Bottom left, Diagram showing how the angle \( \phi \) is measured. Bottom right, Diagram showing the construction of the angle \( \rho \). PA = pulmonary artery; RVO = right ventricular outflow tract; other abbreviations as in figure 1.
of Fallot, the mean value of angle \( \phi \) was 59.2 degrees (range 45 to 80 degrees), whereas in normal subjects it was 23.4 degrees (range 10 to 39 degrees).

Figure 3 shows an example of tetralogy of Fallot with the various measurements and calculations recorded in this case. In figure 4 are shown the data recorded in a control subject.

**Discussion**

The echocardiographic criteria of true dextroposition have not been envisaged as yet. In this report, we attempted to clarify this point using two-dimensional echocardiography to assess the spatial position of the aortic root in 22 patients with tetralogy of Fallot compared with 23 control subjects.

The values of percent rightward displacement (d/a) of the aortic root and especially the position of the crossing-point \( r \) in relation to the aortic cusps show that the aorta is abnormally displaced to the right in relation to the atrial septum in tetralogy of Fallot when compared with normal subjects. The change in the relationships between the atrial septum and the aortic root determined by the position of point \( r \) confirms the real rightward displacement of the aorta in patients with tetralogy of Fallot but does not indicate the responsible mechanism. Indeed, the aortic root can be displaced to the right (dextroposed) as a result of three possible mechanisms (figure 5): (1) rightward translation, which we can speculate to be secondary to the lack of left migration of the conus from an embryologic point of view, (2) clockwise rotation (looking downstream) about an eccentric axis, which we can suppose to be secondary to the lack of conal rotation, or (3) a combination of both mechanisms. The constant position of point \( z \) (anterior aortic commissure), as assessed by the identical value of angle \( \phi \) in patients with tetralogy and in normal subjects, excludes the possibility of a rightward shift of the aorta by a translation; should the aorta be rightward translated with or without additional rotation, then the angle \( \phi \) would be diminished and the point \( z \) displaced to the right (figure 5). The significant diminution of the angle \( \theta \) in patients with tetralogy proves the clockwise rotation of the aortic root. The pivot of the rotation is the anterior aortic commissure as shown by the absence of difference between the value of angle \( \phi \) in the two groups...
TABLE 2
Clinical data and echocardiographic measurements in normal subjects

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex</th>
<th>Age</th>
<th>d/a (°)</th>
<th>r</th>
<th>θ</th>
<th>o</th>
<th>φ</th>
<th>ρ</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>20 yr</td>
<td>82</td>
<td>18</td>
<td>Middle of NCC</td>
<td>+42</td>
<td>R ½-L ½ RCC</td>
<td>16</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>6 mo</td>
<td>90</td>
<td>31</td>
<td>Middle of NCC</td>
<td>+30</td>
<td>R ½-L ½ RCC</td>
<td>14</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>7 yr</td>
<td>87</td>
<td>25</td>
<td>Middle of NCC</td>
<td>+48</td>
<td>R ½-L ½ RCC</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>2 yr</td>
<td>83</td>
<td>17</td>
<td>Middle of NCC</td>
<td>+30</td>
<td>R ½-L ½ RCC</td>
<td>17</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>11 yr</td>
<td>96</td>
<td>11</td>
<td>Middle of NCC</td>
<td>+33</td>
<td>Ant. com.</td>
<td>16</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>27 yr</td>
<td>93</td>
<td>26</td>
<td>Middle of NCC</td>
<td>+43</td>
<td>Ant. com.</td>
<td>13</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>1 yr</td>
<td>87</td>
<td>22</td>
<td>R ½ NCC</td>
<td>+48</td>
<td>R ½-L ½ RCC</td>
<td>13</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>9 yr</td>
<td>76</td>
<td>18</td>
<td>R ½ NCC</td>
<td>+57</td>
<td>R ½-L ½ RCC</td>
<td>15</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>8 yr</td>
<td>80</td>
<td>39</td>
<td>Middle of NCC</td>
<td>+40</td>
<td>L ½ RCC</td>
<td>8</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>5 yr</td>
<td>90</td>
<td>7</td>
<td>Middle of NCC</td>
<td>+30</td>
<td>R ½-L ½ RCC</td>
<td>15</td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>9 yr</td>
<td>95</td>
<td>24</td>
<td>Middle of NCC</td>
<td>+43</td>
<td>Ant. com.</td>
<td>12</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>11 yr</td>
<td>80</td>
<td>33</td>
<td>Middle of NCC</td>
<td>+60</td>
<td>R ½-L ½ RCC</td>
<td>14</td>
</tr>
<tr>
<td>13</td>
<td>F</td>
<td>7 yr</td>
<td>88</td>
<td>28</td>
<td>R ½ NCC</td>
<td>+55</td>
<td>Ant. com.</td>
<td>8</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>1 mo</td>
<td>89</td>
<td>35</td>
<td>Middle of NCC</td>
<td>+42</td>
<td>R ½-L ½ RCC</td>
<td>12</td>
</tr>
<tr>
<td>15</td>
<td>M</td>
<td>5 yr</td>
<td>93</td>
<td>21</td>
<td>Middle of NCC</td>
<td>+37</td>
<td>Ant. com.</td>
<td>13</td>
</tr>
<tr>
<td>16</td>
<td>M</td>
<td>8 yr</td>
<td>94</td>
<td>25</td>
<td>Middle of NCC</td>
<td>+50</td>
<td>Ant. com.</td>
<td>13</td>
</tr>
<tr>
<td>17</td>
<td>M</td>
<td>1 yr</td>
<td>80</td>
<td>15</td>
<td>R ½ NCC</td>
<td>+58</td>
<td>R ½-L ½ RCC</td>
<td>13</td>
</tr>
<tr>
<td>18</td>
<td>F</td>
<td>2 yr</td>
<td>85</td>
<td>31</td>
<td>Middle of NCC</td>
<td>+43</td>
<td></td>
<td>9</td>
</tr>
<tr>
<td>19</td>
<td>M</td>
<td>7 yr</td>
<td>90</td>
<td>24</td>
<td>Middle of NCC</td>
<td>+36</td>
<td>R ½-L ½ RCC</td>
<td>17</td>
</tr>
<tr>
<td>20</td>
<td>M</td>
<td>8 yr</td>
<td>86</td>
<td>19</td>
<td>Middle of NCC</td>
<td>+49</td>
<td>R ½-L ½ RCC</td>
<td>10</td>
</tr>
<tr>
<td>21</td>
<td>F</td>
<td>10 yr</td>
<td>80</td>
<td>17</td>
<td>Middle of NCC</td>
<td>+39</td>
<td>R ½-L ½ RCC</td>
<td>15</td>
</tr>
<tr>
<td>22</td>
<td>M</td>
<td>6 yr</td>
<td>90</td>
<td>32</td>
<td>Middle of NCC</td>
<td>+43</td>
<td>R ½-L ½ RCC</td>
<td>12</td>
</tr>
<tr>
<td>23</td>
<td>M</td>
<td>11 yr</td>
<td>102</td>
<td>25</td>
<td>Middle of NCC</td>
<td>+40</td>
<td>R ½-L ½ RCC</td>
<td>15</td>
</tr>
</tbody>
</table>

Angles ε, θ, φ, and ρ expressed in degrees. Abbreviations as in Table 1.

(table continues)

(figures 5, panel 2) and by the similar relationships of the aortic root with the pulmonary valve (position of the point o) in patients with tetralogy as in control patients. Thus the various measurements performed in patients with tetralogy of Fallot show that the aorta is abnormally displaced to the right because of a clockwise rotation about an eccentric pivot, the aortic anterior commissure. All these abnormalities of the spatial position of the aortic root can only be the result of a true anomaly of embryogenesis. The dilatation of the aorta above a perimembranous ventricular septal defect as a result of abnormal hemodynamics and creating an exaggeration of the normal overriding would not change the spatial position of the aortic root in terms of position of point r and value of angle θ (figure 6).

Our echocardiographic results are in agreement with findings in the morphologic studies conducted by Becker et al.11 and Goor and associates,4,5 who performed virtually identical geometric measurements on postmortem specimens. We agree with these workers that tetralogy of Fallot is characterized by the morphogenetic features of lack of conal rotation and conal malposition. The conal malposition that participates in the abnormal overriding of the aorta is shown anatomically by the anterior deviation of the conal septum, of which the septal insertion is in front of the trabecula septomarginalis.18 This latter anomaly has been previously studied by two-dimensional echocardiography, which permits diagnosis and quantitation of the deviation of the infundibular septum.39 However, the concept of conotruncal malposition as the sole morphogenetic mechanism in tetralogy6,20-23 cannot explain all the features of the condition, since it alone cannot produce the rightward shift of the aorta with clockwise rotation. This latter feature can be explained only by a lack of the normal conal rotation24 that brings the aorta to a dextroposterior position during the norm-

TABLE 3
Statistical analysis with Student’s t test

<table>
<thead>
<tr>
<th>Measurements</th>
<th>Fallot (n = 22)</th>
<th>Normal (n = 23)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angle of atrial septum (c)</td>
<td>87 ± 6.1</td>
<td>86 ± 6.1</td>
<td>NS</td>
</tr>
<tr>
<td>Percent right displacement of the aortic root (d/a)</td>
<td>55.5 ± 9</td>
<td>23.6 ± 7.6</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Angle θ</td>
<td>9.2 ± 11.2</td>
<td>43.3 ± 8.8</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Angle φ</td>
<td>13.2 ± 2.9</td>
<td>13 ± 2.6</td>
<td>NS</td>
</tr>
<tr>
<td>Angle ρ</td>
<td>59.2 ± 10.7</td>
<td>23.4 ± 8.3</td>
<td>&lt;.001</td>
</tr>
</tbody>
</table>

Angles expressed in degrees. All values are expressed as mean ± SD.
mal embryogenesis.25 Using two-dimensional echocardiography to determine the value of the angle ρ as defined above in our patients with tetralogy of Fallot, we were able to quantitate the degree of the clockwise rotation compared with that in normal subjects; in patients with tetralogy, the mean value of ρ was 59.2 degrees (range 45 to 80 degrees), whereas in control subjects it was 23.4 degrees (range 10 to 39 degrees).

Thus, the average degree of the clockwise rotation in patients with tetralogy of Fallot was $59.2 - 23.4 = 35.8$, with extremes of $45 - 39 = 6$ (minimal clockwise rotation) to $80 - 10 = 70$ (major clockwise rotation).

We conclude from this study that in patients with tetralogy of Fallot, there is a true dextroposition of the aorta with a rightward displacement of the aortic root.

FIGURE 3. Left, Two-dimensional echocardiogram from a patient with tetralogy of Fallot (patient 2). Right, Line drawing showing the different measurements and calculations recorded in this patient: $d/a = 0.50$; $r =$ posterior commissure; $o$ is located at the left third of the right coronary cusp; $\theta = 0$ degrees; $\phi = 13$ degrees; $\rho = 75$ degrees. Note the appearance of the left coronary artery (white arrow). RV = right ventricle; other abbreviations as in figures 1 and 2.

FIGURE 4. Left, Two-dimensional echocardiogram in the parasternal short-axis plane at aortic valve level from a normal subject (patient 11). Right, Line drawing showing the different measurements and calculations recorded in this subject. $d/a = 0.24$; $r =$ middle of the noncoronary cusp; $o$ is located at the anterior commissure; $\theta = +43$ degrees; $\phi = 12$ degrees; $\rho = 30$ degrees. Abbreviations as in figures 1 through 3.
due to a clockwise rotation (looking downstream) of the aorta. We believe that this clockwise rotation of the aorta is secondary to the lack of conal rotation. The effects of this morphogenetic mechanism that can be quantitated by two-dimensional echocardiography are not botted out by corrective surgery. This study shows that two-dimensional echocardiography, by means of geometric measurements, may contribute to the eluci-

FIGURE 5. Diagrammatic representations of the different hypothetical mechanisms responsible for dextroposition of aorta. The double-lined circle represents the position of the aortic root after interference of the presumed mechanism. 1. Rightward displaced aortic root caused by a simple translation corresponding to the lack of left migration of the conus. This mechanism should involve a rightward shift of the anterior commissure (point z) with a diminution of the angle $\phi (\phi_2 < \phi_1$; here $\phi_2 = 0$) and a constant value of the angle $\theta (\theta_2 = \theta_1$). 2. Dextroposition of the aorta caused by a clockwise rotation of the aortic root as assessed by the diminution of angle $\theta (\theta_2 < \theta_1$). The axis of the rotation is the anterior commissure (point z) as assessed by the invariability of the angle $\phi (\phi_2 = \phi_1; Z_2 = Z_1$). 3. Dextroposed aorta caused by both mechanisms (translation with rotation), involving a rightward shift of point z and a diminution of angle $\phi (\phi_2 < \phi_1$). Only the mechanism illustrated in the middle panel is in agreement with the echocardiographic findings in our patients with tetralogy of Fallot. \(A = \text{anterior}; \ L = \text{left}; \ P = \text{posterior}; \ R = \text{right.$} \)

FIGURE 6. Summary of the various types of overriding of the aorta. In tetralogy of Fallot, the aorta overrides the right ventricle due to three mechanisms: (1) dilatation of the aorta and leftward shift of the ventricular septum caused by abnormal hemodynamics, which simply increases the normal overriding; (2) anterior deviation of the infundibular septum, which pulls the right coronary leaflet above the right ventricle; (3) rightward displacement of the aortic root (dextroposition) due to a clockwise rotation. Only the second and third mechanisms are the results of true anomalies of embryogenesis. The first mechanism is of functional origin. \(LV = \text{left ventricle}; \ RV = \text{right ventricle}; \ VS = \text{ventricular septum}; \ VSD = \text{ventricular septal defect}$. *Simple increase in the normal overriding.
PATHOPHYSIOLOGY AND NATURAL HISTORY—CONGENITAL HEART DISEASE

dation of the difficult embryologic mechanisms of congenital heart disease, providing the basis for further prospective studies to assess other conotruncal malformations. Indeed, the geometric approach to cardiac anatomy originated by Van Praagh et al.,26 and used on postmortem specimens4, 11, 26–29 can now be performed with two-dimensional echocardiography, which provides the advantages of a safe, noninvasive, and non-limiting method.

We gratefully acknowledge the secretarial assistance of Evelyne Marcilly and Marie-Odile Marchal. We thank Régine Chenu, Evelyne Fontaine, and Daniel Fernandez for their technical assistance.

References


20. Von Rokitansky KF: Die defekte der Scheidewände des Herzens. Vienna, 1875, Braumüller
22. De la Cruz MV, Da Rocha JP: An ontogenetic theory for the explanation of congenital malformations involving the truncus and conus. Am Heart J 51: 782, 1956

Vol. 73, No. 5, May 1986

899
Is the aorta truly dextroposed in tetralogy of Fallot? A two-dimensional echocardiographic answer.

K Isaaz, J L Cloez, F Marçon, A M Worms and C Pernot

Circulation. 1986;73:892-899
doi: 10.1161/01.CIR.73.5.892

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1986 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/73/5/892