Deductive Echocardiographic Analysis in Infants with Congenital Heart Disease

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SUMMARY
A step-by-step approach to cardiac diagnosis utilizing a chest X-ray and echocardiography is described and used to study a normal infant and six infant patients with angiocardio graphically-proven complex congenital heart malformations.

The heart is divided into three major anatomical segments in order to localize the atrial and ventricular chambers and determine the relationship of the great arteries. The atrial chambers are localized by noting the position of the liver on the X-ray. The right atrium is on the same side as the liver with few exceptions. The ventricular chambers are localized by echocardiographically identifying the tricuspid and mitral valves. They are a part of the morphologically right and left ventricles, respectively. As a general rule, the atroventricular valve whose anterior leaflet is continuous with the posterior margin of a semilunar valve is the mitral valve. The atroventricular valve whose anterior leaflet is not continuous with a posterior semilunar valve margin is the tricuspid valve. When the tricuspid valve is to the right of the mitral valve, the ventricles are in their normal positions (ventricular d-loop); when it is to the left, the ventricles are inverted (ventricular l-loop). The relationship of the great arteries is determined by echocardiographically identifying the semilunar valves and noting their positions relative to each other, the interventricular septum and the ventricular chambers. In a ventricular d-loop, the aortic valve is usually to the right of the pulmonary valve. In a ventricular l-loop, the opposite is true. As much as a 17% error may exist when transposition of the great arteries is a component of the malformation. Therefore, as additional aids in semilunar valve identification the R-C intervals of the valves (interval between R wave of electrocardiogram and valve closure on echogram) and outside diameters of the valve roots are measured. Usually the R-C interval is longer and, in the absence of pulmonary stenosis, the outside diameter greater for the pulmonary valve. The segmental diagnoses are then combined to obtain the "type" of heart, and a careful search is made for associated defects.

Using this approach, the positions of the atrial and ventricular chambers and the relationship of the great arteries were accurately determined in the six complex cardiac malformations studied.

Additional Indexing Words:
Cardiac segmental diagnosis
Ultrasound cardiography
Noninvasive techniques
Transposition of the great arteries

In an earlier report the atroventricular and semilunar valves were used as ultrasonic landmarks to obtain an echocardiographic profile of the heart and its great arteries in the normal neonate. Since that study, additional uses of the atroventricular and semilunar valves have resulted in our being able to predict the character of the internal cardiac anatomy in the infant. Echocardiographically, the tricuspid valve can be differentiated from the mitral valve and the pulmonary valve from the aortic valve. By applying the segmental approach to cardiac diagnosis advocated by Van Praagh, it is possible to localize the atrial and ventricular chambers and determine the relationship of the great arteries. In this report a step-by-step approach to cardiac diagnosis utilizing a plain chest X-ray and echocardiography is described and used to study a normal infant and six infants with proven complex congenital heart anomalies.

Methods
Segmental Approach to Cardiac Diagnosis

In the segmental approach to cardiac diagnosis the position of the heart in the thorax is described as being either left-sided (levocardia) or right-sided (dextrocardia). The position of the heart can be determined from either a

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Supported by grant 253 from the Department of Health, Education and Welfare Division of Maternal and Child Health Services, and Research Grants from the Louisville and Jefferson County Heart Associations and the Kentucky Heart Association, and by the WHAS Crusade for Children.

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Received July 15, 1973; revision accepted for publication July 18, 1974.
plain chest roentgenogram or from the echocardiographic location of the valves, i.e., whether they are found to the left or right of the midsternal line.

According to Van Praagh, there are at least eight cardiac segments (separate developmental units) comprising the heart.4 For diagnostic purposes they can be combined to form the following three major cardiac segments: (1) viscerovascularisitus (sinus venosus, common pulmonary vein and primitive atrium); (2) ventricular loop (atrioventricular canal, ventricle and proximal bulbus cordis); and (3) conotruncus (distal bulbus cordis and truncus). The segmental approach to cardiac diagnosis consists of the evaluation and diagnosis of these three segments (fig. 1).

**Viscerovascular Situs: Atrial Localization**

At present, the atrial chambers cannot be identified directly by echocardiography. However, they can be located by noting the position (situs) of the liver on a plain chest film because a constant relationship is usually found between the greater lobe of the liver, suprahepatic portion of the inferior vena cava, right lung and right atrium.2-5 There are three types of situs: solitus, inversus and ambiguous (symmetricus) (fig. 1).6 When the liver is in its normal position on the right side (situs solitus), the atrial chambers are in their normal positions. When it is on the left side (situs inversus), the right atrium is left-sided and the morphologically left atrium is right-sided. When the position of the liver is not clearly

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**Figure 1**

Major cardiac segments (from bottom to top): (1) viscerovascularisitus; (2) cardiac loops (ventricular loop); and (3) relationships between the great arteries (conotruncus). Cardiac tube and great arteries viewed from in front; semilunar and atrioventricular valves, from above; S, superior; I, inferior; L, left; Sp, spleen; TA, truncus arteriosus; BC, bulbus cordis; V, ventricle; RV, right ventricle; LV, left ventricle; A, atrium; RA, right atrium; LA, left atrium. (Reprinted with permission from PAUL MH: Transposition of the great arteries. In Heart Disease in Infants, Children and Adolescents, edited by MOSS AJ, ADAMS FH. Baltimore, Williams & Wilkins, 1968.)

Circulation, Volume 50, December 1974
defined, as is the case with the asplenia and polysplenia syndromes, the situs is ambiguous (uncertain or indeterminate) and the visceral atrial rule does not hold.

Ventricular Loop: Identification of Atrioventricular Valves, Localization of Ventricular Chambers and Identification of Interventricular Septum

The ventricular chambers can be identified directly using echocardiography, by identifying the atrioventricular valves. The atrioventricular valves are formed predominantly from the atrioventricular canal with portions of the mitral valve originating from the ventricle (future morphologically left ventricle) and portions of the tricuspid valve originating from the proximal bulbus cordis (future morphologically right ventricle). In the embryonic development of the heart, if the straight heart tube folds to the right (dextro- or d-loop), the right ventricle and tricuspid valve come to lie anterior and/or to the right of the left ventricle and mitral valve. If it folds to the left (levo- or l-loop), the right ventricle and tricuspid valve come to lie posterior and/or left-sided with respect to the left ventricle and mitral valve. Regardless of whether one is dealing with a levocardia or dextrocardia, if the tricuspid valve is found to the right of and/or anterior to the mitral valve, there has been ventricular d-looping. If it is found to the left of and/or posterior to the mitral valve, there has been ventricular l-looping.

The anterior and posterior leaflets of the mitral and tricuspid valves can be recorded by echocardiography. The mitral valve cannot be differentiated from the tricuspid valve on the basis of their echographic valve patterns, because they are identical. However, the mitral valve can be differentiated from the tricuspid valve through their relationships with the semilunar valves.

In the normal anatomical condition, there is mitral-aortic valve fibrous continuity, i.e., the tissue of the anterior leaflet of the mitral valve at its base is continuous with the tissue of the posterior and left cusps of the aortic valve and the aortic valve ring (fig. 2). Similarity, in the transposed state, with infrequent exceptions, there is mitral-pulmonary valve fibrous continuity. Thus, mitral-semilunar valve fibrous continuity exists in both cases. The echocardiographic technique used to assess mitral-semilunar valve fibrous continuity has been described previously. The echocardiographic evidence of the anatomic relationship is the finding that the echo from the anterior mitral valve leaflet is continuous with the echoes from the posterior margin of the aortic valve (normally related great arteries) or pulmonary valve (transposed great arteries).

To illustrate this crucial point we offer the following two examples. Figure 3 was obtained from a normal subject. The transducer was positioned and pivoted as described previously, so that the sonic beam was moved superiorly and medially along the tissue of the anterior mitral valve leaflet to its base which anatomically is continuous with the tissue of the posterior and left aortic valve cusps and aortic valve ring (mitral-aortic valve continuity). In this example the echo from the anterior leaflet of the mitral valve, at the beginning of systole, is at the same depth as the echoes from the anterior or posterior margin of the aortic valve. Figure 4 was obtained from an angiographically-proven case of d-transposition of the great arteries with an associated large interventricular septal defect. In figure 4A the sonic beam was again moved superiorly and medially along the tissue of the anterior mitral valve leaflet onto the transposed pulmonary valve. Note that at the beginning of systole, differing from the previous situation, the echo from that portion of the anterior mitral valve leaflet which is located in the left ventricle appears to be somewhat more posterior than the echoes from the posterior margin of the pulmonary valve. However, as the sonic beam is moved up the mitral valve leaflet to its base, the echo from the leaflet is seen to ascend to, and be continuous with, the echoes from the posterior margin of the pulmonary valve (mitral-pulmonary valve continuity).

Anatomically, with some exceptions, the tissue of the anterior leaflet of the tricuspid valve is not continuous at its base with either the aortic or pulmonary valve. Viewed from the right atrium, in figure 5A, the anterior leaflet of the tricuspid valve can be seen to be attached to the anterior lateral margin of the right fibrous ring. Looking from the right ventricular aspect (fig. 5B), with valve closure, the anterior tricuspid valve leaflet would move posteriorly toward the interventricular septum as it moved away from the anterior wall of the right ventricle. Most of the interventricular septum is muscular. A small area immediately below the right and posterior aortic valve cusps is thin and membranous. The membranous portion of the interventricular septum is in continuity with the tissue of the aortic wall (normally related great arteries) or pulmonary wall (transposed great arteries). It is important to realize that the total cardiac septum shows a complex longitudinal twist and does not lie in any single plane. The echocardiographic demonstration of this anatomic condition depends on the direction of the sonic beam, the spatial position of the heart in the thorax and the internal cardiac anatomy. Depending upon these factors, the echo from the anterior tricuspid valve leaflet at the onset of systole may appear to be at the...
Figure 3
Echocardiograms in a normal subject showing: (1) the anatomic fibrous continuity between the anterior mitral valve leaflet and the posterior margin of the aortic valve; and (2) the relationship of the anterior tricuspid valve leaflet to the aortic valve root.

Figure 4
Echocardiograms in a patient with d-transposition of the great arteries and large interventricular septal defect showing: (A) the equivalent of the anatomic fibrous continuity between the anterior mitral valve (MVa) leaflet and posterior margin of the transposed pulmonary valve; and (B) the relationship of the anterior tricuspid valve (TVa) leaflet to the pulmonary valve root.
Photographs of a normal heart showing: (A) the attachment of the anterior leaflet of the tricuspid valve to the right fibrous ring as viewed from the right atrium (the septal leaflet has been mislabeled as the posterior leaflet); and (B) the relationship of the anterior leaflet of the tricuspid valve to the right ventricular structures as viewed from the right ventricle.

same depth as the echoes from the anterior, the side or the posterior margin of the aorta (normally related great arteries) or pulmonary artery (transposed great arteries). However, it should be emphasized that even when the echo from the anterior tricuspid valve leaflet appears to be at the same depth as the echoes from the posterior margin of the semilunar valve, it does not mean that it is continuous with them. To illustrate these points we return to our previous two examples. In figure 3 after rotating the sonic beam off the anterior mitral valve leaflet onto the aortic valve, the transducer was further pivoted so that the sonic beam was moved to the right and slightly inferiorly onto the tricuspid valve. Note that the echo from the anterior tricuspid valve leaflet at the beginning of systole appears to be located at the level of the echoes from the anterior margin of the aorta. In figure 4B the sonic beam was again moved to the right and slightly inferiorly off the pulmonary valve onto the anterior leaflet of the tricuspid valve. Occasionally in the presence of a large interventricular septal defect the echo from the anterior tricuspid valve leaflet may appear to be at the same depth as the echoes from the posterior margin of the aortic valve (normally related great arteries) or, as in this case, the posterior margin of the transposed pulmonary valve. Note that at the position of the arrow the echo from the anterior tricuspid valve leaflet and the echoes from the posterior margin of the pulmonary valve are clearly separated and, therefore, not continuous with each other. It might be argued that the cycle before the one identified can be interpreted as showing a continuous relationship between the anterior tricuspid valve leaflet echo and the echoes from the posterior margin of the pulmonary valve since the echoes are not separated. The difference between the echographic tracings in the two cycles can be accounted for through a small variation in beam angulation. However, the important thing to be stressed is that their simultaneous presence actually precludes a diagnosis of the type of atrioventricular-semilunar valve continuity which exists in the case of the mitral valve. Anatomically, as well as echographically, the anterior leaflet of the mitral valve and the posterior margin of the semilunar valve are not present at the same time. When one ends, the other begins. Thus the anterior leaflet of the tricuspid valve is not continuous with the posterior margin of the aortic valve in the presence of normally related great arteries, and it is not continuous with the posterior margin of the pulmonary valve in the presence of transposition of the great arteries.

What relationship does the tricuspid valve bear to the other semilunar valve? In the normal condition they are separated by an outflow tract (subpulmonary conus). Similarly, in complete transposition of the great arteries they are separated by a subaortic conus. The echocardiographic evidence of this type of anatomical relationship is demonstrated in figure 6 which was obtained from the normal subject studied later in this paper. The transducer was placed in the tricuspid valve recording position and the sonic beam was directed through its anterior leaflet. The transducer was then pivoted so that the sonic beam was moved superiorly off the anterior tricuspid valve leaflet (anterior right atrioventricular valve) onto an anterior outflow tract. In order to record the pulmonary valve

Figure 5

Example 1. Echocardiographic tracing in a normal heart showing the relationship of the anterior leaflet of the tricuspid valve (anterior, right AV valve) to the aortic valve (post., inferior, right SL valve) root and the pulmonary valve (ant., superior left SL valve) root.

Figure 6
DEDUCTIVE ECHO ANALYSIS IN INFANTS

Conotruncus: Identification of Semilunar Valves, Localization of Outflow Tract(s) and Determination of Relationship of Great Arteries

Conal muscular development (growth, lack of growth, and absorption),[4,18] beneath the semilunar valves is one of the most important factors in the embryonic morphogenesis of normal and abnormal relationships between the great arteries and ventricles. It is largely responsible for variations in semilunar valve heights (inferior-superior relationships) and for variations in semilunar valve antero-posterior relationships. The presence of conal tissue below a semilunar valve prevents atrioventricular-semilunar valve fibrous continuity and it lifts the semilunar valve to an anterior, superior position. The absence of conal tissue permits atrioventricular-semilunar valve fibrous continuity and the semilunar valve remains posterior and inferior. Anatomically, there are four possible types of conus: (1) subpulmonary; (2) subaortic; (3) bilaterally present (subpulmonary and subaortic); and (4) bilaterally absent.

Echocardiographically, the presence of an anterior superior semilunar valve which is not in direct continuity with an atrioventricular valve indicates a conus below the semilunar valve (fig. 6: subpulmonary conus below the anterior superior left-sided pulmonary valve). The finding of a posterior inferior semilunar valve which is in direct continuity with an atrioventricular valve precludes the existence of a conus below the semilunar valve (fig. 3: absent conus beneath the aortic valve; and fig. 4A: absent conus below the transposed pulmonary valve).

The truncus gives rise to the semilunar valves and great artery roots. In most infants it is possible to record both semilunar valves from directly overhead. When obtained in this manner, the echographic patterns are identical and cannot be used to differentiate between them. However, in most cases it is possible to differentiate between the aortic and pulmonary valves by analyzing the following: (1) the right-left spatial relationship between the cardiac valves in the frontal plane; (2) the length of the intervals measured between the R waves of the electrocardiograms, which serve as reference points, and the valve closures on the echograms (R-C intervals); (3) the outside diameters of the valve roots.

Ventricular looping is a second major factor in the embryonic morphogenesis of the arterio-ventricular relationships.[3,4,18] It is the primary determinant of the right-left spatial relationship between the semilunar valves. With ventricular d-looping, the aortic valve comes to lie to the right of the pulmonary valve. With ventricular l-looping, it is found to the left of the pulmonary valve. This arterioventricular relationship is known as the loop rule.2 Once the mitral and tricuspid valves have been identified, the loop rule can be used to predict the identity of the semilunar valves. If the tricuspid valve is found to the right of the mitral valve (ventricular d-loop), the aortic valve should be the right-sided semilunar valve. If it is found to the left of the mitral valve (ventricular l-loop), the aortic valve should be the left-sided semilunar valve. In addition, the aortic and pulmonary valves have been identified they can be used to predict the locations of the ventricular chambers.

In many instances an accurate identification of the semilunar valves can be made by comparing their R-C intervals. As long as the pulmonary resistance is lower than the systemic resistance, which is usually the case even in the neonate and infant, the pulmonary valve will close after the aortic valve and the R-C interval will be longer for the pulmonary valve.20

The measurement of the outside diameters of the semilunar valve roots is also very helpful in suggesting the identity of the aortic and pulmonary valves. Normally the outside diameter of the pulmonary valve root is larger than that of the aortic valve root.21,31

Once the semilunar valves have been identified, they are used to determine the relationship of the great arteries. This is done by analyzing their positions relative to each other, the interventricular septum and the ventricular chambers. According to the terminology advocated by Van Praagh,23 if the aorta normally arises from the left ventricle and the pulmonary artery arises normally from the right ventricle, the great arteries are normally related to the ventricles. If the aorta originates from the right ventricle and the pulmonary artery from the left ventricle, the great arteries are transposed. If they both arise from the right ventricle (double outlet right ventricle),23,24 the left ventricle (double outlet left ventricle)25 or a single ventricle,26 the great arteries are said to be malpositioned. The prefix d- is used when the aortic valve is to the right of the pulmonary valve; the prefix l-, when it is to the left. If the aortic valve is neither to the right nor to the left of the pulmonary valve, i.e., it is either directly anterior or posterior to it, the prefix A- or P- should be used to denote that relationship.

Integration of Segments

Once the cardiac position and the anatomic diagnosis of each segment is established, the various diagnoses are integrated to represent the over-all picture or "type" of heart.4

Associated Defects

Following the delineation of the "type" of heart, a search is made for associated defects within individual segments and between adjacent segments: defects such as a malattachment of the anterior leaflet of an atrioventricular valve, stenosis of an atrioventricular or semilunar valve and intra- or extracardiac shunts. This is accomplished by: (1) noting the relationships between the atrioventricular and semilunar valves; (2) noting the relationship between the interventricular septum and semilunar valves; (3) analyzing the qualitative motion of the cardiac valves and interventricular septum; and (4) analyzing the quantitative measurements of the cardiac valves, cardiac chambers, heart walls and great arteries.

Echocardiographic Techniques

Echocardiographic examinations were performed as described previously1 with a commercially available ultra-
sonoscope (Smith Kline Ekoline 20) utilizing a 5 MHz, 8
mm diameter nonfocused transducer (Aerotech Labs) and a
repetition rate of 1,000 impulses/sec. All echograms were
recorded directly from the oscilloscope screen onto Polaroid
film or on a strip-chart recorder (Electronics for Medicine
DR-8). Measurements of pertinent cardiac structures were
made according to the methods described in our normal
study.3

Patients
A 14-day-old, 3.75 kg, white male infant, who was judged
to have a normal heart on the basis of a normal history,
physical examination, electrocardiogram and plain chest
film, and six patients with complex congenital cardiac
malformations were studied by echocardiography utilizing
the approach described above. All six patients were also
fully studied by cardiac catheterization and angiocardi-
diography.

Results

Example 1
Normal 3.75 kg, 14-day-old white male infant.

Cardiac Position and Visceroatrial Situs

Examination of the plain chest film revealed the
heart to be on the left side (levocardia) and the liver to
be on the right (situs solitus).

Echocardiographic Valve Positions

Looking at figure 7A, all four valves were found to
lie to the left of the midsternal line. The atroventricular
valves were found below the 3rd and 4th interspaces;
one just adjacent to the sternum (posterior left
atrioventricular valve) and the other anterior and to
the right of this position under the left border of the
sternum (anterior right atrioventricular valve). A
posterior semilunar valve was found under the inferior
border of the 3rd rib at its junction with the sternum
(posterior inferior right semilunar valve) and an
anterior semilunar valve was found superior to and to
the left of this position under the 2nd left interspace
(anterior superior left semilunar valve).

Ventricular Loop

Rotation of the sonic beam off the anterior leaflet of
the posterior left atrioventricular valve showed its
echo to be continuous with the echoes from the
posterior margin of the posterior inferior right
semilunar valve (fig. 8). Rotation off the anterior
leaflet of the anterior right atrioventricular valve
failed to show any continuous relationship with the
posterior margin of any semilunar valve (fig. 6).
Therefore, it follows that the posterior left atroventricular
valve is the mitral valve and the anterior right
atrioventricular valve is the tricuspid valve. Since the
tricuspid valve lies to the right of the mitral valve, it is
a ventricular d-loop (fig. 7B).

An interventricular septum with normal motion
(anterior during ventricular diastole and posterior
during ventricular systole)27 was found separating the
anterior right ventricle from the posterior left ventri-
cle (fig. 8).

Conotruncus

In a ventricular d-loop the aortic valve should lie to
the right of the pulmonary valve. If the loop rule
holds, the right semilunar valve which is posterior,
inferior and in direct continuity with the mitral valve
is the aortic valve, the left semilunar valve which is
anterior, superior and has an outflow tract between it
and the right ventricle is the pulmonary valve and the
great arteries are d-normally related to the ventricles.
To confirm whether or not the loop rule holds,
the R-C intervals of the semilunar valves were
measured. In figure 9 the semilunar valves were
serially recorded by manually moving the transducer,
and thereby the sonic beam, superiorly and leftward off the posterior inferior right semilunar valve onto the anterior superior left semilunar valve. Note that the R-C intervals of the anterior and posterior semilunar valves are 0.24 sec and 0.22 sec, respectively. Since the R-C interval is longer for the anterior semilunar valve, the anterior superior left semilunar valve is the pulmonary valve, the posterior inferior right semilunar valve is the aortic valve, the loop rule holds and the great arteries are d-normally related to the ventricles.

The diagnoses of the cardiac position and cardiac segments in this subject are: levocardia, situs solitus, ventricular d-loop and d-normally related great arteries (fig. 7C).

Integration of Segments

Situs solitus means that the right atrium is right-sided. Ventricular d-loop means that the right ventricle is right-sided. D-normally related great arteries means that the aortic valve is to the right of the pulmonary valve, and that the aorta arises from the left ventricle and the pulmonary artery from the right ventricle. Thus, a right-sided right atrium communicates with a right-sided right ventricle which gives rise to an anterior superior left-sided pulmonary artery through an outflow tract (subpulmonary conus). A left-sided left atrium communicates with a left-sided left ventricle which gives rise directly to a posterior inferior right-sided aorta (mitral-aortic valve continuity). This is a normal "type" heart.

Associated Defects

Qualitatively, the atrioventricular-semilunar valve relationships, interventricular septal-semilunar valve relationship and echographic movements of the atrioventricular valves, semilunar valves and interventricular septum were normal. Quantitatively, the atrioventricular and semilunar valve mobilities, chamber dimensions, wall thicknesses and great
arterial root dimensions were within normal limits.

Thus, this subject had a normal anatomical echocardiographic study, i.e., a normal “type” heart plus a normal qualitative and quantitative echocardiographic profile.

Example 2

A 5-day-old, 3.045 kg, cyanotic white male.

Cardiac Position and Visceroatrial Situs

Examination of the plain chest roentgenogram (fig. 10A) shows the heart to be on the left side (levocardia) and the liver to be on the right (situs solitus).

Echocardiographic Valve Positions

Looking at figure 11A, a posterior left atrioventricular valve, an anterior right atrioventricular valve, a posterior inferior left semilunar valve and an anterior superior right semilunar valve are found to the left of the midsternal line.

Ventricular Loop

Looking at figure 12A, the anterior leaflet of the posterior left atrioventricular valve is continuous with the posterior margin of the posterior inferior left semilunar valve. In figure 12B the anterior leaflet of the anterior right atrioventricular valve is not continuous with the posterior margin of either semilunar valve. Thus, the posterior left atrioventricular valve is the mitral valve, the anterior right atrioventricular valve is the tricuspid valve and it is a ventricular d-loop (fig. 11B).

An interventricular septum with normal septal motion is seen in figure 12A.

Conotruncus

In a ventricular d-loop the right-sided semilunar valve, which in this case is anterior, superior and separated from the right ventricle by an outflow tract (fig. 12B), should be the aortic valve. Similarly, the left-sided semilunar valve, which in this case is inferior, posterior and in direct continuity with the mitral valve (fig. 12A), should be the pulmonary valve. Looking at figure 12C, the R-C interval of the posterior inferior left semilunar valve is longer than that of the anterior superior right semilunar valve. Therefore, the anterior superior right semilunar valve is the aortic valve and the posterior inferior left semilunar valve is the pulmonary valve.

Figure 10

Plain chest films showing: (A) a left-sided heart and a right-sided liver (levocardia, situs solitus); (B) a left-sided heart and liver (levocardia, situs inversus); and (C) a right-sided heart and liver (dextrocardia, situs solitus).

Diagnosis:

Levoicardia Situs Solitus

Figure 11

Example 2. Frontal view drawing showing: (A) the locations of the cardiac valves; (B) the labeling of the atrioventricular (AV) valves and ventricular loop following the echocardiographic identification of the AV valves; and (C) the labeling of the semilunar (SL) valves and relationship of the great arteries following the echocardiographic identification of the SL valves.
semilunar valve is the pulmonary valve. The loop rule holds and there is d-transposition of the great arteries.

The diagnoses of the cardiac position and cardiac segments in this patient are: levocardia, situs solitus, ventricular d-loop and d-transposition of the great arteries (fig. 11C).

Integration of Segments

Situs solitus means that the right atrium is right-sided. Ventricular d-loop means that the right ventricle is right-sided. D-transposition of the great arteries means that a right-sided aorta arises from the right ventricle and a left-sided pulmonary artery from the left ventricle.

Thus, a right-sided right atrium communicates with a right-sided right ventricle which gives rise to an anterior superior right-sided aorta through an outflow tract (subaortic conus). A left-sided left atrium communicates with a left-sided left ventricle which gives rise directly to a posterior inferior left-sided pulmonary artery (mitral-pulmonary valve continuity).

Figure 12

Example 2: Echocardiograms showing: (A) posterior left AV valve-posterior inferior left SL valve continuity and normal septal motion; (B) anterior right AV valve-SL valve discontinuity; and (C) a longer R-C interval for the posterior inferior left SL valve and a larger outside diameter for the posterior artery.

Circulation, Volume 50, December 1974
Associated Defects

Analysis of the qualitative and quantitative echocardiographic profiles revealed no evidence of valvular stenosis. In addition, no conclusions could be drawn as to the presence or absence of intra- or extracardiac shunts.

Angiocardiography

In figure 13 (A-1 & B-1) the position of the catheter as it enters the heart on the right side confirms the location of the inferior vena cava as assumed from the plain chest film (situs solitus). Comparing A-1 to B-1, the right ventricle lies to the right of the left ventricle (ventricular d-loop). Looking at the angiograms, an anterior right-sided aorta arises from the right ventricle by way of an outflow tract (d-transposition of the great arteries). The posterior left-sided pulmonary artery originates from the left ventricle. In addition, there is an interventricular septal defect and an angiographically documented patent ductus arteriosus.

Example 3

A 3-month-old, 4.827 kg, acyanotic white female.

Cardiac Position and Visceroatrial Situs

Examination of the chest film (fig. 10B) reveals the heart and liver to be on the left side (levocardia, situs inversus).

Echocardiographic Valve Positions

The positions of the cardiac valves were very similar to those found in example 2 (fig. 11A). The major difference was that the atrioventricular valves were found lying more in a side-by-side position than an antero-posterior position.

Ventricular Loop

In a manner similar to example 2 (fig. 12A & 12B), the posterior left atrioventricular valve was shown to be the mitral valve, the anterior right atrioventricular valve was shown to be the tricuspid valve and it was proved to be a ventricular d-loop.

An interventricular septum with paradoxical motion (posterior during ventricular diastole and anterior during ventricular systole) was recorded.

Conotruncus

Applying the loop rule, the right-sided semilunar valve should be the aortic-valve, the left-sided semilunar valve should be the pulmonary valve and the great arteries should be d-transposed to the ventricles. In a manner similar to example 2 (fig. 12C), comparison of the R-C intervals confirmed that the right-sided semilunar valve was the aortic valve, the left-sided semilunar valve was the pulmonary valve and there was d-transposition of the great arteries.

The diagnoses of the cardiac position and cardiac segments in this patient are: levocardia, situs inversus, ventricular d-loop and d-transposition of the great arteries.

Integration of Segments

The diagnoses in this patient are similar to those of the preceding patient except for the visceroatrial situs which results in a right-sided left atrium (situs inversus) communicating with a right-sided right ventricle (ventricular d-loop). As in the previous example, the right ventricle gives rise through an outflow tract to an anterior right-sided aorta (d-transposition of the great arteries). The left-sided right atrium communicates with a left-sided left ventricle from which a posterior left-sided pulmonary artery originates.

Associated Defects

Analysis of the qualitative and quantitative echocardiographic profiles revealed paradoxical septal motion and a thick anterior right ventricular wall. This suggested the presence of an interventricular septal defect, patent ductus arteriosus or tricuspid valve regurgitation with right ventricular volume overload.

Angiocardiography

In figure 14 (A-1 & B-1) the position of the catheter as it enters the heart on the left side confirms the presence of a left-sided inferior vena cava (situs inversus). Looking at A-1 and B-1, the ventricles are lying
side-by-side with the right ventricle being right-sided (ventricular d-loop). Looking at the angiograms, an anterior right-sided aorta arises from the right ventricle by way of an outflow tract (d-transposition of the great arteries). The posterior left-sided pulmonary artery originates from the left ventricle. In A-1 there is evidence of an intra-cardiac shunt. Analysis of the catheterization data revealed it to be at the ventricular level.

Example 4
A 2-year-old, 10 kg, acyanotic white male.

Cardiac Position and Visceroatrial Situs
The plain chest film revealed levocardia and situs solitus.

Echocardiographic Valve Positions
The positions of the cardiac valves are depicted in figure 15A: a posterior left atrioventricular valve; an anterior right atrioventricular valve; a posterior inferior right semilunar valve; and an anterior superior left semilunar valve.

Ventricular Loop
The anterior leaflet of the posterior left atrioventricular valve is not continuous with the posterior margin of either semilunar valve. In figure 16A the anterior leaflet of the anterior right atrioventricular valve is continuous with the posterior margin of the posterior inferior right semilunar valve. Therefore, the anterior right atrioventricular valve is the mitral valve and the posterior left atrioventricular valve is the tricuspid valve. Since the tricuspid valve is to the left of the mitral valve, it is a ventricular l-loop (fig. 15B).

An interventricular septum with paradoxical motion was recorded (not shown).

Conotruncus
In a ventricular l-loop, the left-sided semilunar valve, which in this case is anterior, superior and separated from the left-sided right ventricle by an outflow tract should be the aortic valve. Similarly, the right-sided semilunar valve, which is posterior, inferior and in direct continuity with the mitral valve, should be the pulmonary valve. Since in figure 16B the R-C interval of the posterior inferior right semilunar valve is longer than that of the anterior

![Figure 14](image1.png)

Example 3. Angiocardiograms showing: the left-sided inferior vena cava (situs inversus); the right-sided morphologically right ventricle (ventricular d-loop); the morphologically right ventricle (RV) giving rise to the right-sided aorta (Ao) and the morphologically left ventricle (LV) giving rise to the pulmonary artery (d-transposition of the great arteries); and an intra-cardiac shunt.

![Figure 15](image2.png)

Figure 15
Example 4. Frontal view drawing showing: (A) the locations of the cardiac valves; (B) the labeling of the atrioventricular (AV) valves and ventricular loop following the echocardiographic identification of the AV valves; and (C) the labeling of the semilunar (SL) valves and relationship of the great arteries following the echocardiographic identification of the SL valves.
superior left semilunar valve, the posterior inferior right semilunar valve is the pulmonary valve and the anterior superior semilunar valve is the aortic valve. The loop rule holds and there is l-transposition of the great arteries.

The diagnoses of the cardiac position and cardiac segments in this patient are: levocardia, situs solitus, ventricular l-loop and l-transposition of the great arteries (fig. 15C).

**Integration of Segments**

A right-sided right atrium (situs solitus) communicates with a right-sided left ventricle which gives rise to a right-sided pulmonary artery. A left-sided left atrium communicates with a left-sided right ventricle (ventricular l-loop) which gives rise by way of an outflow tract to a left-sided aorta (l-transposition of the great arteries).

**Associated Defects**

Analysis of the qualitative and quantitative echocardiographic profiles revealed paradoxical motion of the ventricular septum and a markedly enlarged left atrial cavity diameter. The unusually large size of the left atrium suggested the presence of either a large left-to-right shunt or left atrioventricular valve regurgitation.

**Angiocardiography**

In figure 17 (A-1) the catheter enters the heart on the right (situs solitus). Looking at A-1 and B-1, the right ventricle tends to be posterior, superior and left-sided with respect to the left ventricle (ventricular l-loop). Looking at the angiograms, the right-sided left ventricle gives rise to a posterior right-sided pulmonary artery and the left-sided right ventricle gives rise by way of an outflow tract to an anterior left-sided aorta (l-transposition of the great arteries). In B-1 and B-2 there is evidence of a marked left-sided tricuspid valve regurgitation.

**Example 5**

A 1½-year-old, 10.34 kg, cyanotic white female.

**Cardiac Position and Visceroatrial Situs**

The chest film revealed levocardia and situs solitus.

**Echocardiographic Valve Positions**

The positions of the cardiac valves were practically identical to those found in the preceding example (fig. 15A).
superior left semilunar valve. Thus, the posterior inferior right semilunar valve is the pulmonary valve and not the aortic valve. The anterior superior left semilunar valve is the aortic valve and not the pulmonary valve. The loop rule does not hold and there is l-transposition of the great arteries.

The diagnoses of the cardiac position and cardiac segments in this patient are: levocardia, situs solitus, ventricular d-loop and l-transposition of the great arteries.

**Integration of Segments**

A right-sided right atrium (situs solitus) communicates with a right-sided right ventricle (ventricular d-loop) which gives rise to a left-sided anterior aorta through an outflow tract (l-transposition of the great arteries). A left-sided left atrium communicates with a left-sided left ventricle from which a right-sided posterior pulmonary artery originates.

**Associated Defects**

Qualitatively, when the sonic beam is moved along the interventricular septum onto the transposed pulmonary artery (fig. 18A) there is a consistent drop out of the interventricular septal echotrace suggesting the presence of a large septal defect. Quantitatively, since the outside diameter of the pulmonary root is only 9 mm and the R-C interval of the pulmonary valve is 0.38 sec compared to 0.30 sec for the aortic valve (fig. 18C), it follows that there is pulmonary stenosis.

**Angiocardiography**

In figure 19 (A-1 & B-1) the catheter enters the heart on the right (situs solitus). In the lateral view (A-2 & B-2) the right ventricle is anterior to the left ventricle. In the frontal view (A-1 & B-1) the ventricles are superimposed. However, the positions of the catheters indicate that the tricuspid valve is to the right of the mitral valve (ventricular d-loop). Thus, a right-sided right atrium communicates with an anterior right ventricle and a left-sided left atrium communicates with a posterior left ventricle. Looking at the angiograms, the anterior right ventricle gives rise through an outflow tract to an anterior left-sided aorta (l-transposition of the great arteries). The posterior left ventricle gives rise to a posterior right-sided pulmonary artery. In B-1 and B-2 there is angiographic evidence of an interventricular septal defect and pulmonary stenosis.

**Example 6**

A 4-day-old, 2.84 kg, cyanotic white male.

**Cardiac Position and Visceroatrial Situs**

Analysis of the chest film was consistent with levocardia and situs solitus.
Echocardiographic Valve Positions

The positions of the cardiac valves are illustrated in figure 20A. A posterior left atrioventricular valve and an anterior right atrioventricular valve are found in their usual positions. In contrast to the first four examples, two anterior superior semilunar valves are found lying side-by-side at the level of the 2nd interspace.

Ventricular Loop

In figure 21A the anterior leaflet of the posterior left atrioventricular valve is related to the posterior margin of a left-sided outflow tract. By manually moving the transducer from the 3rd interspace to the 2nd interspace it is possible to move the sonic beam up the outflow tract to the anterior superior left semilunar valve which is recognized through the movements of its valve leaflets. In figure 21B the anterior leaflet of the anterior right atrioventricular valve is not continuous with the posterior margin of any structure. By manually moving the transducer and sonic beam up the right-sided outflow tract, it is possible to record the anterior superior right semilunar valve. Since neither atrioventricular valve has an anterior leaflet that is in direct continuity with the posterior margin of a semilunar valve, the mitral and tricuspid valves cannot be identified directly.

There was no echocardiographic evidence of an interventricular septum. The diagnoses to this point are as shown in figure 20B.

Conotruncus

In figure 21C the R-C interval of the anterior superior left semilunar valve is longer than that of the

Figure 18

Example 5. Echocardiograms showing: (A) posterior left AV valve-posterior inferior right SL valve continuity and the dropping out of echoes from the interventricular septum; (B) anterior right AV valve-SL valve discontinuity; and (C) a longer R-C interval for the posterior inferior right SL valve and an outside diameter of 9 mm for the posterior artery root.
anterior superior right semilunar valve. In addition, the outside diameter of the left-sided arterial root is larger than that of the right-sided one. Thus, the anterior superior left semilunar valve is the pulmonary valve and the anterior superior right semilunar valve is the aortic valve (fig. 20C). If the loop rule holds, the posterior left atrioventricular valve is the mitral valve and the anterior right atrioventricular valve is the tricuspid valve.

The diagnoses of the cardiac position and cardiac segments in this patient are: levocardia, situs solitus, single ventricle, bilateral conus and d-malposition of the great arteries (fig. 20C).

Integration of Segments

A right-sided right atrium (situs solitus) communicates with a single ventricular chamber through what is probably a tricuspid valve. A left-sided left atrium communicates with the ventricular chamber through what is probably a mitral valve. From the ventricular chamber a bilateral (subaortic and subpulmonary) conus connects to an anterior right-sided aorta and an anterior left-sided pulmonary artery (d-malposition of the great arteries).

Example 5. Angiocardiograms showing: the right-sided inferior vena cava (situs solitus); the anterior morphologically right ventricle (ventricular d-loop); the morphologically right ventricle (RV) giving rise to the left-sided aorta (Ao) and the morphologically left ventricle (LV) giving rise to the pulmonary artery (l-transposition of the great arteries); and an interventricular septal defect and pulmonary stenosis.

Figure 19

Example 6. Frontal view drawing showing: (A) the locations of the cardiac valves; (B) the labeling of single ventricle and bilateral conus following the echocardiographic examination of atrioventricular-semilunar valve relationships and the ventricular segment; and (C) the labeling of the semilunar (SL) valves, and by application of the loop rule, the atrioventricular (AV) valves, and the determination of the relationship of the great arteries following the echocardiographic identification of the SL valves.

Figure 20

Associated Defects

Analysis of the qualitative and quantitative echocardiographic profiles provided no evidence of any additional defects.

Angiocardiography

In figure 22 (A-1 & B-1) the catheter enters the heart on the right (situs solitus). Looking at the angiograms, from a single ventricular chamber two outflow tracts (bilateral conus) can be seen leading to a right-sided aorta and a left-sided pulmonary artery (d-malposition of the great arteries).

Example 7

A 1-day-old, 2.045 kg, acyanotic white female.

Cardiac Position and Visceroatrial Situs

Examining figure 10C, the heart is on the right side (dextrocardia) and the liver is on the right (situs solitus).
Figure 21
Example 6. Echocardiograms showing: (A) posterior left AV valve-anterior superior left SL valve discontinuity; (B) anterior right AV valve-anterior superior right SL valve discontinuity; and (C) a larger outside diameter for the anterior left artery.

Echocardiographic Valve Positions

Figure 23A depicts the positions of the cardiac valves. All four valves are found to the right of the mid-sternal line: a posterior left atrioventricular valve; an anterior right atrioventricular valve; a posterior inferior right semilunar valve; and an anterior superior left semilunar valve.

Ventricular Loop

The posterior left atrioventricular valve was shown to be the mitral valve by the demonstration of atrioventricular-semilunar valve continuity, the anterior right atrioventricular valve was shown to be the tricuspid valve and it was proved to be a ventricular d-loop (fig. 23B).

An interventricular septum with normal septal motion was recorded.

Conotruncus

If the loop rule holds, the right-sided semilunar valve which is inferior, posterior and in continuity with the mitral valve is the aortic valve; the left-sided semilunar valve, which is anterior, superior and separated from the right ventricle by an outflow tract is the pulmonary valve; and the great arteries are d-normally related to the ventricles. Comparison of the R-C intervals confirmed that the right-sided semilunar valve was the aortic valve, the left-sided semilunar valve was the pulmonary valve and there were d-normally related great arteries.
Example 6. Angiocardiograms showing: the right-sided inferior vena cava (situs solitus); a single ventricular chamber (vent.); and a bilateral conus (not labeled) leading to a right-sided aorta (Ao) and a left-sided pulmonary artery (d-malposition of the great arteries).

The diagnoses of the cardiac position and cardiac segments in this patient are: dextrocardia, situs solitus, ventricular d-loop and d-normally related great arteries (fig. 23C).

Associated Defects

The qualitative echocardiographic profile is within normal limits. Applying the methodology and deductive reasoning presented in a previous report, analysis of the quantitative echocardiographic profile in Table 1 suggests the presence of an interatrial septal defect, interventricular septal defect and patent ductus arteriosus.

Angiocardiography

In Figure 24 the catheter enters the right-sided heart (dextrocardia) on the right (situs solitus). Looking at Figure 24A, the right ventricle is right-sided (ventricular d-loop) and it gives rise to a left-sided pulmonary artery by way of an outflow tract (d-normally related great arteries). In Figure 24B and 24C the left-sided left atrium communicates with a left-sided left ventricle from which a right-sided aorta originates. In addition, there is angiographic evidence of an intracardiac left-to-right shunt. By catheterization there was evidence of an interatrial septal defect, interventricular septal defect and patent ductus arteriosus.

Discussion

The segmental approach to cardiac diagnosis using the terminology advocated by Van Praagh is an ideal systematic approach to the diagnosis and description of congenital heart disease. It is an anatomicoembryological approach which has been evolved from a number of apparent morphogenetic facts. Essentially, one diagnoses the anatomic type of each major cardiac segment and then integrates the various segmental diagnoses to obtain the “type” of heart. A search is then made for associated defects.

The most important step toward the development of echocardiography into a definitive tool for the study of congenital heart disease is the establishment of a simple standardized technique for the ultrasonic examination of the heart and its great arteries. If in the presence of congenital heart disease the anatomy and developmental rotation of the heart remained the same from patient to patient, which it does not do, it would be possible to select a number of precordial locations from which a standardized set of echograms could be obtained. To some extent this has been the approach adopted by the majority of investigators.

In a previous publication on the normal neonate, we described a method which was designed to take advantage of the unique characteristics of the neonate.
Table 1

Echocardiographic Data Form

<table>
<thead>
<tr>
<th>Type of heart</th>
<th>Position in thorax</th>
<th>Visceeroatrial situs</th>
<th>Ventricular loop</th>
<th>Conotrunceus</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>L</td>
<td>Levo cardiac</td>
<td>S</td>
<td>Solitus</td>
</tr>
<tr>
<td></td>
<td>D</td>
<td>Dextrocardia</td>
<td>I</td>
<td>Inversus</td>
</tr>
<tr>
<td></td>
<td>A</td>
<td>Ambiguous</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Comments: Dextrocardia; normal type heart; normal qualitative echocardiographic profile.

Echocardiographic dimensional profile

<table>
<thead>
<tr>
<th>Measurements (mm)</th>
<th>Measurements (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal range*</td>
<td>Infant</td>
</tr>
<tr>
<td>Anterior AV valve mobility</td>
<td>8.5 – 11.7</td>
</tr>
<tr>
<td>AP diameter of ant vent</td>
<td>10.1 – 14.3</td>
</tr>
<tr>
<td>Thickness of ant vent wall</td>
<td>1.0 – 2.8</td>
</tr>
<tr>
<td>Outside diameter ant artery</td>
<td>10.4 – 12.5</td>
</tr>
<tr>
<td>Anterior SL valve opening</td>
<td>5.5 – 7.3</td>
</tr>
<tr>
<td>AP diameter of LA cavity</td>
<td>6.5 – 10.2</td>
</tr>
<tr>
<td>Posterior AV valve mobility</td>
<td>8.3 – 10.9</td>
</tr>
<tr>
<td>AP diameter of post vent</td>
<td>15.8 – 21.4</td>
</tr>
<tr>
<td>Thickness of septum</td>
<td>2.0 – 3.2</td>
</tr>
<tr>
<td>Thickness of post vent wall</td>
<td>1.9 – 3.3</td>
</tr>
<tr>
<td>Outside diameter post artery</td>
<td>9.0 – 11.0</td>
</tr>
<tr>
<td>Posterior SL valve opening</td>
<td>3.9 – 5.3</td>
</tr>
</tbody>
</table>

Interpretation: Echocardiographic study compatible with left-to-right shunting at atrial, ventricular and ductus levels.

*For normal values see reference 2.

and infant, i.e., a cartilagenous skeletal system over the tricuspid valve and a lack of intervening lung tissue over the pulmonary valve. Since the echographic patterns of the atrioventricular and semilunar valves were easy to recognize, the valves were used as ultrasonic cardiac landmarks. The transducer was placed on the precordium directly above each of the valves and the sonic beam was directed anteroposteriorly through the valve. In a study of 240 normal newborns, the precordial locations directly under the valves were found remained relatively constant from subject to subject because the internal locations of the atrioventricular and semilunar valves remained relatively constant. What about the valve locations in the presence of congenital heart disease? In many congenital heart defects the internal valve locations are essentially the same as those of the normal subject, e.g., in valvular aortic stenosis. In some, the valve locations are slightly altered, e.g., the alteration of the semilunar valve locations in d-transposition of the great arteries. In a few, such as the dextrocardias, the valve locations are markedly altered. Thus, depending upon the internal location of the valves, the precordial valve recording positions in the patient with congenital heart disease may essentially be the same as those of a normal subject or may, as in the case of dextrocardia, be markedly altered. Therefore, the first step in the echocardiographic examination of any patient with a congenital heart defect is the determination of the locations of the atrioventricular and semilunar valves. Once they have been located, they can be identified and used as ultrasonic landmarks to investigate the heart and its great arteries.

The echographic recognition of atrioventricular-semilunar valve continuity and discontinuity is the key to the identification of the atrioventricular valves and subsequent localization of the ventricular chambers. Gramiak and Shah were the first to delineate the echographic equivalent of the anatomic fibrous continuity existing between the anterior leaflet of the mitral valve and the posterior and left cusps of the aortic valve and the aortic valve ring. In the example selected for publication the sonic beam was moved superiorly and medially along the tissue of the anterior mitral valve leaflet onto the aortic valve. The echo from the anterior mitral valve leaflet recorded in the left ventricle, left ventricular outflow tract and mitral-aortic valve junction at the beginning of systole were all found at the same depth as the echoes from the posterior margin of the aortic valve. At the mitral-aortic valve junction an arrow was placed to show that the echo from the anterior mitral valve leaflet ended and the echoes from the posterior margin of the aortic valve began without a break in the echotrace. Chesler et al. applied the technique developed by Gramiak.
and his associates to examine the mitral-semilunar valve relationships of a number of patients with proven congenital heart anomalies. Echographic mitral-aortic valve continuity was found in patients with tetralogy of Fallot, pseudotruncus arteriosus, interventricular septal defects and dextrocardia with normally related great arteries. Echographic mitral-pulmonary valve continuity was found to be present in patients with both d- and l-transposition of the great arteries. In five patients with double outlet right ventricle in whom mitral-semilunar valve continuity was not present angiographically, the echo from the anterior mitral valve leaflet at the onset of systole was found to lie 10 mm or more posterior to the echoes from the posterior margin of the semilunar valves. In the example shown, the anterior mitral valve leaflet echo and the echoes from the posterior margin of the semilunar valve were clearly discontinuous, i.e., there was a break (gap) in the echotrace. However, so much emphasis was placed on the antero-posterior discrepancy found between the echoes of the anterior mitral valve leaflet and semilunar valve, that confusion has arisen over what constitutes echographic mitral-semilunar valve discontinuity. In our opinion, the antero-posterior relationship between the echo of the anterior mitral valve leaflet recorded in the left ventricle or left ventricular outflow tract and the echoes from the posterior margin of the semilunar valve has nothing to do with whether mitral-semilunar valve discontinuity is present or not. If the sonic beam is moved superiorly and medially along the tissue of the anterior mitral valve leaflet onto the semilunar valve, and the anterior mitral valve leaflet echo ascends to, and is continuous with, the echoes of the posterior margin of the semilunar valve, then there is continuity. For echographic mitral-semilunar valve discontinuity to be present, there must be a consistent break in the echotrace, or there must be echoes from an intervening structure separating the anterior mitral valve leaflet from the echoes of the posterior margin of the semilunar valve.

The identification of the semilunar valves and subsequent localization of the great arteries rests upon the analysis of the right-left spatial relationship between the cardiac valves in the frontal plane, the length of the R-C intervals of the semilunar valves and/or the size of the valve roots. From anatomic studies, the loop rule would be expected to hold in any congenital heart defect in which transposition of the great arteries is not a component. Transposition of the great arteries was not a component in 93% of 10,624 cases seen at the children’s hospital in Boston.29 It was a component in 7%. Exceptions to the loop rule have occurred in anatomic (postmortem) studies with the following frequencies: (1) dextrocardia, 4% (2/51 cases); (2) single (common) ventricle, 8% (5/60 cases); and (3) transposition of the great arteries of many types, 5% (7/149 cases).30 Angiocardiographically, Guerin et al.31 found, in a study of transposition of the great arteries of many types, that the loop rule failed in 9% (6/70 cases) and Carr et al.45 found in a study confined to classical complete transposition that it failed in 17% (19/109 cases).

To ascertain whether or not the loop rule holds, the R-C intervals of the semilunar valves are measured and compared. Following birth, the expansion and ventilation of the lungs is associated with a dramatic fall in the pulmonary vascular resistance, and the elimination of the very low resistance placental
vascular bed is associated with a marked increase in the systemic vascular resistance. When the resistance of the pulmonary vascular bed falls below that of the systemic vascular bed, the pulmonary valve comes to close after the aortic valve. We studied, at sea level, 40 normal newborns during the first few hours and days of life by echocardiography. We found, at the first measurement in room air, that the R-C interval of the pulmonary valve was longer than that of the aortic valve in 80% and equal to it in 20%. By placing the subjects in whom we were unable to measure a difference in 100% oxygen for 15 minutes, we were able to increase the number in whom the pulmonary valve R-C interval was longer to 95%.

We have analyzed the echograms of 160 infants with various forms of congenital heart disease proved by cardiac catheterization and angiography (unpublished data). Because we only recently began recording the semilunar valves on a strip chart recorder, we cannot evaluate their R-C intervals. However, we do have phonocardiograms with simultaneous pressure tracings obtained at the time of catheterization on 159 of them. Of these, 147 had two semilunar valves and 138 had split second heart sounds with the pulmonary component occurring after the aortic component. Thus, if the R-C interval had been available for measurement it would have been longer for the pulmonary valve in at least 94% of the infants studied with two semilunar valves. None of the nine infants in whom splitting was not appreciated were placed in 100% oxygen for 15 minutes, which may have increased the percentage. Of the 147 infants studied with two semilunar valves, 30 had systolic pressures in the pulmonary artery of systemic levels (systolic pressure in the pulmonary artery within 10% of the systolic pressure in the aorta). Despite these high systolic pressure levels, the second heart sound was documented to be split in 70% of them. In the patients in whom splitting was appreciated, the diastolic pressure in the pulmonary artery was noted to be lower than the diastolic pressure in the aorta.

Factors which can be expected to influence pulmonary vascular changes after birth include hypoxia and acidemia. Hypoxia, due to alveolar hypoventilation or exposure to low oxygen in the environment after birth, may result in a marked delay in the fall of the pulmonary vascular resistance. It has been shown that children born and living at higher altitudes have higher pulmonary vascular resistances and pulmonary arterial pressures than those at sea level. Similar findings have also been reported in a group of infants who were hypoxic due to lung disease or hypoventilation from cerebral causes. In these patients the second heart sound may not be split.

The measurement of the outside diameters of the great arterial roots becomes important when the R-C intervals of the semilunar valves either cannot be adequately determined, or if determined, are found to be equal. In two large studies on the normal neonate, the outside diameter of the pulmonary root was found to be larger than that of the aortic root. These echocardiographic findings are consistent with measurements made at autopsy on a series of normal children's hearts. In the 147 infants with congenital heart disease studied by us, in whom there were two semilunar valves, the pulmonary root was found to be larger than the aortic root in 86%. If the patients with pulmonary stenosis are excluded, it was larger than the aortic root in 95%. Exceptions were found in two patients with simple d-transposition of the great arteries, two patients with left-to-right shunts, and one patient with pulmonary vein atresia and a reversed ductus. The exceptions in the patients with d-transposition of the great arteries are not surprising in light of the fact that Shaher found the pulmonary and aortic root sizes to be equal in 54% (57/105 cases) of those transpositions in which there was no interventricular septal defect or pulmonary stenosis. The reason for the exceptions in the other three patients is not readily apparent.

Let us now turn our attention to the cases with congenital heart disease. In all six of our angiographically proven cases, the location of the inferior vena cava, and by inference the right atrium, was accurately determined from the position of the liver on the plain chest film. While this almost always will be the case, partial visceroatrial discordance has been known to occur. As an example, Hastreiter and Rodriguez-Coronel reported three cases in which the liver and right atrium were found on opposite sides of the midline. In these cases the right lung was found on the same side as the right atrium and the inferior vena cava switched sides at the level of the liver to drain into the right atrium. Thus, these rare cases had thoracic visceroatrial concordance (right lung-right atrial concordance) and venoatrial concordance (inferior vena cava-right atrial concordance), with abdominal visceroatrial discordance (hepatic-right atrial discordance). With rare exceptions, the inferior vena cava drains into the right atrium even though it may have to switch sides to do so. As an alternate method to using the abdominal viscera to localize the atrial chambers, Van Mierop et al. have proposed using the tracheobronchial tree to identify the right and left lungs. The routine study of the tracheobronchial tree may have some value because of the occasional finding of partial visceroatrial discordance and because according to Van Mierop and his associates, bilateral right main bronchi (bilateral right lungs) are pathognomonic of the asplenia syndrome and bi-
lateral left main bronchi (bilateral left lungs) strongly favor the polysplenia syndrome. Recently, we have begun to incorporate this into our approach. However, our experience is too limited to draw any conclusions.

Examples 2-5 and 7 support the assumption that the ventricular chambers can be identified with absolute certainty when an interventricular septum and two atrioventricular valves are present, if the anterior leaflet of one atrioventricular valve (mitral valve) is continuous with the posterior margin of a semilunar valve and the anterior leaflet of the other atrioventricular valve (tricuspid valve) is not. When the tricuspid valve is anterior and to the right of the mitral valve, angiocardiographically, the right ventricle is usually located anterior and to the right of the left ventricle. Rarely, as in example 5, it may be situated directly anterior to the left ventricle. When the tricuspid valve is found directly to the right of the mitral valve, the right and left ventricles tend to lie side-by-side. In contrast to the usual antero-posterior relationship found between the ventricular chambers in a ventricular d-loop, in a ventricular l-loop, the ventricular chambers are usually found lying side-by-side and/or superior-inferior to each other. Rarely, exceptions can occur, as in the case in example 6, where there is bilateral atrioventricular-semilunar valve discontinuity. In this situation the loop rule, which is an indirect method, can be used to predict the morphology of the atrioventricular valves. In this example, since the aortic valve was determined to be the right-sided semilunar valve, the anterior right atrioventricular valve was labeled the tricuspid valve and the posterior left atrioventricular valve was labeled the mitral valve. Since both atrioventricular valves appeared to open into a single ventricular chamber, the morphology of the chamber could not be specified.*

In the six examples selected, the loop rule accurately predicted the locations of the atrioventricular valves and ventricular chambers in five. In example 5 the loop rule failed because of conotruncal-ventricular discordance. This example brings out the importance of using the R-C intervals and/or the outside diameters of the great arterial roots to confirm or deny the validity of the loop rule. If only one atrioventricular valve is recorded, then there are four possibilities: (1) the second atrioventricular valve has not been recorded; (2) it is a common atrioventricular valve; (3) there is tricuspid atresia; or (4) there is mitral atresia. In our experience it has been rare that we have had trouble locating two atrioventricular valves when they have been present. When the problem has occurred, it has almost without exception been in the patients with a ventricular l-loop where the ventricular chambers were found lying side-by-side and/or superior-inferior to each other. In our hands the echographic pattern obtained from a common atrioventricular valve has usually been different from that obtained from a normal atrioventricular valve. When only one normal atrioventricular valve is present, if its anterior leaflet is continuous with the posterior margin of a semilunar valve, then with possible rare exceptions, it is the mitral valve, and the ventricular chamber into which it opens is the left ventricle (tricuspid atresia). If its anterior leaflet is not continuous with the posterior margin of either semilunar valve, then in all probability it is the tricuspid valve and the ventricular chamber into which it opens is the right ventricle (mitral atresia).

The recognition of the echoes from the interventricular septum by Edler et al. and their subsequent use by Popp et al. to study the motion of the interventricular septum, as well as to estimate the size of the right and left ventricular cavities, represents one of the most significant advances in the development of echocardiography. As a cardiac structure the interventricular septum is one of the most difficult to study. Even the use of angiocardiography has led to erroneous conclusions, because of artifacts. In contrast to the angiocardiographic method, if one echocardiographically records a septum, it is there. The problem arises when the septum is not found. In example 5 no septum was recorded. Since in this patient the atrioventricular valves were definitely situated antero-posterior to each other, so that the interventricular septum would be expected to lie in a plane favorable to the reflection of ultrasound, the absence of an echographic septum was considered probable proof of a single ventricle. On the other hand, if the atrioventricular valves had been found lying side-by-side at the same depth below the transducer, as is commonly the case in a ventricular l-loop, then we would have been hesitant to make such a diagnosis. In this situation the interventricular septum would be expected to lie somewhat parallel to the ultrasonic beam path. Since the amount of ultrasound which is reflected from an interface is minimized when the angle of incidence of the sound beam to the reflecting surface approaches 180 degrees, it is not surprising that such a septum may not be recorded. In examples 3 and 4 the interventricular septum was recorded, but only after considerable difficulty. Before concluding

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*At autopsy there was a single ventricular chamber with the morphology of a right ventricle. A bilateral conus led to a right-sided aorta and a left-sided pulmonary artery. The anterior right atrioventricular valve had the characteristics of a tricuspid valve and the posterior left atrioventricular valve had the characteristics of a mitral valve.
that an echographic septum is not present, the patient should be rotated to the left and to the right, as well as progressively moved from a supine position to a sitting position, in order to try and bring the interventricular septum, if present, into a plane favorable to its recording. Of the five remaining patients, two had paradoxical septal motion. In example 3 there was a volume overload on the right ventricle as a result of the interventricular septal defect, i.e., the increased blood flow through the lungs returned to the right-sided right ventricle by way of the right-sided left atrium. In example 4 the mechanism responsible for the paradoxical motion was probably on a different basis from volume overloading of the right ventricle. Since the ventricles were inverted, it would not have been surprising to find that the septum moved anteriorly during ventricular systole as the anterior left ventricle contracted. What is hard to explain in this patient is the existence of paradoxical motion in the presence of the large left-sided tricuspid valve regurgitation, a defect which results in a volume overload on the posterior right ventricle. It should also be mentioned that paradoxical motion may be seen in normal hearts, when the sonic beam is directed through that portion of the interventricular septum which is in the immediate vicinity of the semilunar valve.41

Echocardiography is the only noninvasive method available for the study of atrioventricular-semilunar valve relationships. In addition, it is often superior to angiocardiographic methods. The examples in this paper show that it is possible to demonstrate echocardiographically the presence of interventricular-semilunar valve continuity and discontinuity. As demonstrated in all seven examples, the most superior precordial level below which the atrioventricular valves were recorded remained virtually constant at the level of the 3rd interspace. In contrast, the levels at which the semilunar valves were recorded varied depending upon the presence or absence of a conus. In the absence of a conus there was atrioventricular-semilunar valve continuity and the semilunar valve was found below the 3rd interspace in a posterior position. In the presence of a conus the semilunar valve was recorded in an anterior position at the level of the 2nd interspace. In example 6 there was a bilateral conus and both semilunar valves were found lying side-by-side in anterior positions below the 2nd interspace.

It has been shown throughout this study that it is possible in individual cases to accurately identify the aortic and pulmonary valves by applying the loop rule, by measuring the R-C intervals of the semilunar valves and/or by measuring the outside diameters of the great arterial roots. At the present time, most investigators determine the relationship of the great arteries solely on the basis of the semilunar valve positions.42 43 They do not identify the left and right ventricles, nor do they actually identify the individual semilunar valves. In essence, the law of probabilities is used. Ventricular d-looping is very common; ventricular l-looping is relatively rare. Great arterioventricular concordance is very common; great arterioventricular discordance is rare. Thus, when an anterior semilunar valve is found to the right of a posterior semilunar valve and the statement is made that there is transposition of the great arteries, the probability of being correct is clearly favorable. However, example 5 proves that such an approach will inevitably lead to an occasional erroneous diagnosis. If echocardiography is to become a reliable tool for the study of congenital heart disease, we must try to accurately diagnose each major cardiac structure. To make a diagnosis of transposition of the great arteries we must prove that the pulmonary artery arises from the left ventricle and that the aorta originates from the right ventricle.

The seven examples in this paper with congenital heart disease represent selected cases. Obviously, it is not always possible to obtain sufficient information to make an accurate diagnosis. However, the determination of the atrial and ventricular chambers usually can be accomplished. By applying the loop rule and taking into account whether the patient is cyanotic or acyanotic, it is generally possible to arrive at a good precatheterization diagnosis.

Step-by-Step Approach to the Study of Congenital Heart Disease

The first step in the study of any patient with congenital heart disease, or one suspected of having heart disease, is the examination of the plain chest roentgenogram to ascertain the position of the heart and, if possible, the lungs in the thorax (levocardiography or dextrocardia) and the position of the liver in the abdomen. By deduction, the positions of the liver and lungs determine the locations of the atrial chambers.

The second step is the determination of the number of atrioventricular and semilunar valves present and their locations. This is accomplished by moving the transducer over the precordium until the valves are located. Once they have all been located, the transducer is directed antero-posteriorly through each valve, and the position of the transducer on the precordium is noted. From the position of the transducer on the precordium, the atrioventricular valves are labeled as being right and left and the semilunar valves as being right and left and inferior and superior. By measuring the distance between the transducer and the echographic patterns of the valves on the echogram, they are labeled as being anterior and posterior.
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The third step is to identify the mitral and tricuspid valves and through them determine the positions of the ventricular chambers. This is accomplished by rotating and/or moving the sonic beam off the anterior leaflet of each atrioventricular valve onto the semilunar valves. When the anterior leaflet of one atrioventricular valve is continuous with the posterior margin of a semilunar valve and the anterior leaflet of the other atrioventricular valve is not, the one that is continuous is the mitral valve and the other is the tricuspid valve. If the tricuspid valve lies to the right of the mitral valve, it is a ventricular d-loop. If it lies to the left of the mitral valve, it is a ventricular l-loop.

The fourth step is to determine the echocardiographic presence or absence of an interventricular septum by rotating and/or moving the transducer, and thereby the sonic beam, off the atrioventricular valves onto and across the ventricular chambers.

The fifth step is to identify the aortic and pulmonary valves and through them determine the type(s) of atrioventricular-semilunar valve continuity and/or conus (coni) which is (are) present and the relationship of the great arteries. This is accomplished by noting the positions of the semilunar valves in the frontal plane and applying the loop-rule. To verify that the loop rule holds, the R–C intervals of the semilunar valves are analyzed. If the intervals cannot be obtained, or they are found to be equal, then the outside diameters of the arterial roots are analyzed.

The sixth step is to integrate the segmental diagnoses in order to ascertain the “type” of heart that is present. In essence this determines the basic floor plan of the heart.

The seventh step is to search for associated defects. This is accomplished by analyzing the echocardiographic profile of the patient in terms of the following qualitative parameters: (1) atrioventricular-semilunar valve relationships; (2) interventricular septal-atrioventricular valve relationship; and (3) atrioventricular valve, semilunar valve and interventricular septal movements. In addition, quantitative measurements are made of the cardiac valves, cardiac chambers, heart walls and great arterial roots.

The time required to complete the first six steps is variable. It can often be accomplished in 15 minutes. Rarely, it may take as long as 1 1/2 hours. The primary rate limiting factor is the time spent searching for the anterior superior semilunar valve. For the beginner it is the most difficult valve to locate. Its detection depends upon its anterior position and the recognition of valve leaflet movements. In the last 100 normal newborns studied, our detection rate was 98%. In similar studies Chung et al.44 and Hagan et al.21 reported rates of 92% and 80%, respectively. The presence of a pneumomediastinum, pneumothorax, or severe respiratory distress prevents the recording of the anterior superior semilunar valve. It cannot be recorded when there is intervening air or lung tissue. In the 160 infants with congenital heart disease referred to earlier, we were able to record two semilunar valves in 89%.

How long should one take to do an echocardiographic examination? The clinical state of the patient should dictate the time that can or should be spent in examination. If it is decided that a patient would benefit from emergency cardiac catheterization or surgery, then 15 to 20 minutes seems a reasonable time to spend. If the patient is to remain in the nursery, then once the temperature, electrolytes, pH and oxygen environment have been stabilized and are being monitored, we feel it is safe to take sufficient time to obtain good data.

Acknowledgment

We wish to thank Dr. A. S. Abbasi for his suggestion to document this approach and Mary Adkins, Kathy Pollard, Donna Oiler and Marilyn Jackson for their technical assistance. Dr. Solinger wishes to thank Dr. Alma L. Roby, without whose financial assistance this paper could not have been written.

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Deductive Echocardiographic Analysis in Infants with Congenital Heart Disease
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Circulation. 1974;50:1072-1096
doi: 10.1161/01.CIR.50.6.1072
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1974 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:
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