Echocardiographic Spectrum of Inter-arterial and Arterioventricular Relationship of Great Arteries

KALIM U. AZIZ*  MILTON H. PAUL**
AND ALEXANDER J. MUSTER**

SUMMARY:

Eighteen patients with varied Congenital heart disease were selected from amongst 820 echocardiograms performed between 1978-79 in order to demonstrate echo imaging of inter-arterial and arterio-ventricular relations. Sector scan echocardiograms were attained using an A.T.L. Sector scanner with 3.5 mHz transducer. Echocardiographic imaging of normally related great vessels was correlated with angiograms in 2, double outlet right ventricular connections in three and four patients with transposed great vessel connections. Angiographic, anatomic post mortem and sector scan image correlates were demonstrated in four patients with corrected transposition and four patients with Tetralogy of Fallot. One patient showed echo features of double outlet left Ventricle.

It is concluded that 2D sector scan echocardiography is capable of demonstrating the whole spectrum of ventriculo-arterial and inter-arterial relationship.

The cardiac connections of the great arteries vary greatly in various congenital cardiac malformations. Traditionally, angiography has been the mainstay for the diagnosis of inter-arterial and arterio-ventricular relationships of the great arteries. Recently popularized axial views have improved the diagnostic capability of the angiography (1). Two-dimensional sector scan echocardiography has been shown to provide diagnostic imaging of the normally related and transposed great arteries relationships (2-9). Complicated arterio-ventricular connections such as double outlet right ventricle can be diagnosed by two-dimensional echocardiography (9) and atrioventricular connections can be studied with precision by echocardiography (10). The purpose of the present report is to illustrate a correlative angiographic, post-mortem anatomic and two-dimensional echocardiographic spectrum of inter-arterial and arterio-ventricular relationships in 18 patients.

**Children's Memorial Hospital, Chicago, Illinois U.S.A.
* Pediatric cardiologist NICVD, Karachi.

MATERIAL AND METHODS

Eighteen patients were selected from amongst 820 echocardiograms performed between 1978-1979 on children with various congenital cardiac malformations. These 18 patients comprised 2 patients with normally related great vessels, 4 patients with tetralogy of Fallot, 3 patients with double outlet right ventricle and subpulmonary stenosis. Four patients had d-transposition of the great arteries (simple transposition), 4 had l-transposition (corrected transposition) and 1 patient had double outlet left ventricle. The diagnosis was made prospectively in 14/18 patients. In 4 patients the diagnosis was known prior to echocardiography; 3 of these had l-transposition and 1 had double outlet right ventricle (Table-I). Angiographic confirmation of the echo diagnosis was available in all. Right and left ventricular angiograms in various axial views were obtained in order to establish the interarterial and arterio-ventricular connections.

Two dimensional sector scan and M-mode echocardiograms were obtained in all unsedated patients in supine position, using an ATL sector
TABLE-I: GREAT VESEL ECHO SPECTRUM

<table>
<thead>
<tr>
<th>Number</th>
<th>Age (Years)</th>
<th>Associated Lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Range</td>
<td>V.S.D.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Subpulmonary</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Stenosis</td>
</tr>
<tr>
<td>Normally related great vessels</td>
<td>2</td>
<td>0.6 &amp; 2.0</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>4</td>
<td>0.5 - 3.4</td>
</tr>
<tr>
<td>Double outlet right ventricle</td>
<td>2</td>
<td>0.5 &amp; 2.3</td>
</tr>
<tr>
<td>Taussig-Bing (DORV)</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>d-TGA</td>
<td>4</td>
<td>2 d - 18</td>
</tr>
<tr>
<td>1-TGA</td>
<td>4</td>
<td>0.3 - 26</td>
</tr>
<tr>
<td>Double outlet left ventricle</td>
<td>1</td>
<td>16.0</td>
</tr>
<tr>
<td>Total</td>
<td>18</td>
<td></td>
</tr>
</tbody>
</table>

d = days
V.S.D. = Ventricular Septal defect
DORV = Double outlet right ventricle.
d-TGA = Simple transposition of the great arteries.
1-TGA = Corrected transposition of the great arteries.

scanner with 3.5 mHz transducer. The sector images were recorded on Sanyo videotape and stop frame photographs were obtained with photographic camera. Angiographic and post-mortem anatomic data were correlated. The cardiac images were obtained from various locations (2-5) and in some later studies high left parasternal location was utilized, and high resolution echo images were obtained particularly in small infants and newborns. The transducer was positioned at the 2nd-3rd left intercostal space,

2-3 cm from the sternum, and the sector beam was kept at an oblique angle to the aortic-left ventricular axis (long axis) and directed inferiorly and medially toward the sternum. In this position the left ventricle, aorta and a small portion of the outflow of the right ventricle were visualized, and with leftward and superior tilting of the transducer toward the left shoulder the right ventricular outflow, pulmonary artery and right pulmonary artery coursing behind the ascending aorta could be well visualized (Fig.-1).

![Diagram](image)

FIGURE - 1, I

A, B, Diagrammatic representation of the transducer location and echo beam planes using a high left parasternal location (2nd to 3rd intercostal space 3-4 cm away from the midline). The echo beam transects the left ventricle-aorta long axis in A. Superior and leftward tilting of the transducer images the right ventricular outflow tract, main pulmonary artery and right pulmonary artery above the left atrium (B).
to us when describing the great vessel imaging.

Transducer rotation motion between the planes of section is referred to as counterclockwise and clockwise and transducer tilting motion is referred to as superior, inferior, left or right. The echo images obtained are displayed in a mode as approximate as possible to the orientation of the heart within the body.

RESULTS

Normally related great vessels could be imaged in various planes from varied transducer locations. From the subxiphoid location with sector beam in left ventricular long axis plane, the origin of the ascending aorta from the left ventricle could be imaged. The ascending aorta was shown to continue into the aortic arch and in smaller subjects origins of cranial vessels could be demonstrated. The cross sectional image of the right pulmonary artery was to the left of the ascending aorta in this view (Fig. 2 C). The transducer was then rotated clockwise so that sector beam was eventually brought in the right ventricular sinus-pulmonary valve axis, in approximate sagittal body plane (left ventricular “short-axis” plane). Minor adjustments of rotation and posterior displacement of the transducer were then required to image the right ventricular sinus, ventricular septum, right ventricular outflow, pulmonary valve annulus, and main pulmonary artery in all patients irrespective of age (Fig.2 A-B). In the newborn period the origin of the right and left pulmonary arteries could also be visualized. The right ventricular-pulmonary artery section image was seen in this view as a crescent shape anterior to a circular left ventricle. A rightward and posterior tilting of transducer displayed the aortic image arising from the left ventricle (Fig. 2-D). Thus, from subxiphoid position it was possible to establish the origin of a posterior aorta from the left ventricle and anterior leftward pulmonary artery from the right ventricle.

Confirmation of the normally related great vessel relationship could be obtained in short axis image obtained with transducer located parasternally at the base of the heart. The aorta in this view was seen as a circle anterior to the left atrium. The right ventricular outflow tract was seen anterior to the aorta and curved leftward to join the main pulmonary artery, which bifurcated into the right and left pulmonary arteries (7) (Fig.-3-A). The left ventricular image obtain-

FIGURE — 1, II.

A, Sector scan echocardiogram obtained with transducer position A in Figure 1. Imaging of the left ventricle (LV) and aorta (AO) is well seen. B, Superior and leftward tilting of the transducer from position A images the right ventricular outflow tract (RVO), main pulmonary artery (MPA) and right pulmonary artery (RPA) above the left atrium (LA).

TERMINOLOGY AND IMAGE DISPLAY

The transducer positions were named suprasternal, left parasternal, apical and subxiphoid or substernal. Planes of section of the heart were in reference to the left ventricle; a) long axis, b) short axis, c) four chamber (11).

For suprasternal location the plane of sections were referred to as sagittal and coronal body planes, since reference to long and short left ventricular section planes seems inappropriate.
ed with transducer positioned at high left para-
ternal location with echo beam in left ventricu-
lar long axis plane could also be used to identify
normally related great vessels. The left ventricle
and aorta were visualized in long axis and with
superior tilting of the transducer, the ascending
aorta up to the arch could be imaged; superior
and leftward tilting of the transducer imaged an
anterior leftward pulmonary artery (Fig. 1).

Left ventricular long-axis image obtained
from the standard parasternal location could
also be used to demonstrate normally related
great vessels. With sector beam transecting the
left ventricular long axis anteroposteriorly, the
aorta and left ventricle were imaged, however,
the aortic arch was not well seen in this view; a
clockwise rotation of the transducer with superior
and leftward tilting imaged the right ventricular
outflow tract and the main pulmonary artery.

From the suprasternal location it was possible
to image the great vessels in all patients irrespec-
tive of age. With the sector beam in the coronal

FIGURE 2.

A. Post mortem heart specimen sectioned in the
sagittal body plane. B. Correlative short axis plane sector
scan, and C. Left ventricular long axis planar view obtained
from subxiphoid location in a subject with normally
related great arteries. Origin of the pulmonary artery
from right ventricle (B) and aorta from left ventricle is
seen.

Ao, aorta; LV, left ventricle; MB, muscle bundle;
MPA, main pulmonary artery; PA, pulmonary artery; RV,
right ventricle; S, septum.
A, Cross sectional great vessel view at the base of the heart, obtained from parasternal location in a subject with normally related great arteries, and C, in a patient with 1-transposition of the great arteries. In the patient with normally related great arteries the right ventricular outflow tract (RVO) forms a curvature anterior to a circular aorta (Ao). The main pulmonary artery (PA) and its branches are located to the left of the aorta. In d-transposition the aorta is anterior and to the right of pulmonary artery (PA), B, while aorta is anterior and to the left of pulmonary artery in 1-transposed great arteries (C).

Body plane, the aortic arch was imaged in cross section, superior vena cava to the right in long-axis, and right pulmonary artery was inferior, seen as parallel lines from its origin to the lung hilum (Fig.-4 A-C). Anterior tilting of the transducer beam from this position imaged the ascending aorta up the aortic sinuses with superior vena cava as its right relation (3) (Fig.-4 B,D). Counterclockwise rotation of the transducer from coronal to near sagittal plane was required to bring the echo beam parallel to the aortic arch.

With an anterior tilting of the transducer from this position at times it was possible to image the left pulmonary artery anterior to the descending aorta (3).

The displacement of the aorta above the right ventricle with its partial connections to the right and left ventricles was demonstrated in four patients with tetralogy of Fallot. Subxiphoid location was most useful; with sector beam plane transecting the left ventricular long axis, ascending aorta and arch of the aorta could be imaged.

Overriding of the aorta above the ventricular septal defect could be well visualized (Fig.-5-C). Rotation of the sector beam to left ventricular short-axis plane demonstrated an anterior pulmonary artery arising from a crescent-shaped right ventricle. Infundibular stenosis and subcrystal ventricular septal defect could be well seen (Fig.5-B). Echo contrast studies and anatomic section in sagittal plane confirmed the echocardiographically recognized structures. Overriding of the aorta above the ventricular septum could also be demonstrated in left ventricular long-axis plane obtained at left parasternal position and high parasternal position (Fig.-1).

Origin of both great vessels from the right ventricle (double outlet right ventricle) could be imaged from subxiphoid location in left ventricular long-axis plane. The ascending aorta up to the transverse arch could be seen to the right of the pulmonary artery, and with tilting of the echo beam to the left, the main pulmonary artery and its
FIGURE 5.

A, Lateral view (sagittal plane) cineangiogram in a patient with tetralogy of Fallot, B, Correlative left ventricular short axis planar view echocardiogram obtained from subxiphoid location, and C, Sector scan in ventricular long axis plane from subxiphoid location. Angiogram shows the position of hypertrophied crista (arrows) which is seen as a thick transverse echo in B. The ventricular septal defect (D) lies below the hypertrophied crista supraventricularis. Overriding of the aorta above the ventricular septal defect (D) is well seen in C.

first branch could be imaged (Fig.-6). Muscular subpulmonary stenosis was well seen in all patients. The ventricular septal defect was imaged to be committed to the pulmonary artery in both patients (Fig.-7 A,B).

Double outlet right ventricle of Taussig-Bing type with subpulmonic ventricular septal defect was visualized in left ventricular long-axis plane from parasternal location, which showed an anterior aorta and pulmonary artery overriding the ventricular septum above the ventricular septal defect (Fig.-7 C,D). Severe muscular subpulmonary stenosis was well seen. From the subxiphoid location in left ventricular long-axis plane, the identity of the anterior vessel as the aorta was confirmed by its continuation into the arch located to the right of the pulmonary artery. The extent of the pulmonary arterial override could be precisely evaluated.

In complete transposition of the great arteries (d-TGA), the connections of the aorta to the right ventricle and pulmonary artery with its first branching to the left ventricle could be demonstrated from subxiphoid location in left ventricular long-axis plane in all four patients (8). (Figs. 8 and 9). From suprasternal location an anterior and rightward aorta in transverse section and posterior, leftward main and right pulmonary artery in longitudinal section could be imaged (Fig.-10). From the left parasternal location with the echo beam in short axis plane at the base of the heart, aorta was located anteriorly and to the right of the pulmonary artery (d-transposition) (Fig.-2-B). Our recent experience with high left parasternal location suggests that the diagnostic imaging of transposed vessels can also be obtained. From this position with echo beam oblique to the left ventricular long axis, pulmonary artery left ventricle communication could be imaged and superior tilting of the transducer from this position showed the origin of aorta from the right ventricle (Fig.-11 A,B).

In corrected, or 1-transposition of the great arteries, from subxiphoid location in four chamber plane, the left ventricle, recognized by lack of atrioventricular valve attachments to the ventricular septum and smooth-walled chambers,
A, Right ventricular (four chamber view) angiogram, B, Correlative left ventricular long axis planar view sector scan obtained from subxiphoid location, and C-D, Left ventricular long axis view obtained from parasternal position, in a patient with double outlet right ventricle. Angiogram shows origin of both great arteries from the right ventricle, and pulmonary artery overriding (black arrow) the ventricular septum (S) above the ventricular septal defect (D). Sector scans correlate the anatomic features seen in the angiograms. Note that the precise degree of pulmonary arterial override can be assessed on echocardiogram (B,C). The subpulmonary muscular stenosis is well seen on echocardiograms (white border arrows). Anterior and rightward transducer angulation from position C to D is required to optimally image an anterior aorta.

was right-sided and connected with a right-sided right atrium (Fig.-12-A). Superior tilting of the transducer from four-chamber plane to long axis plane showed a vertical origin of the main pulmonary artery and its branches from the left ventricle in all patients (Figs.-12-A and 13-B). Continued anterior and leftward tilting of the transducer showed aorta arising from a left-sided right ventricle (Fig.-13-A). Horizontal (1-patient) or oblique ventricular septum (3 patients) and left-sided triangular right ventricle with septal attachment of the tricuspid valve, coarse, trabeculated pattern, was imaged with discordant left atrium to right ventricle connections (Fig.-12-B)

and 13-B). In one patient pulmonary artery was atretic and additionally in 3 of 4 moderately large ventricular septal defect was visualized.

Left parasternal short axis plane imaging at the base of the heart showed the aorta sectioned transversely as a circle to the left of a posterior pulmonary artery (Fig.-3). In the suprasternal view in coronal body plane, the anterior aorta was leftward and the pulmonary artery posterior and rightward (Fig.-14 A,B). An anterior leftward tilting of the transducer visualized the section of the ascending aorta up to the aortic valve, located to the left of the pulmonary artery (Fig.-14-A).

The origin of both great vessels from the left ventricle was diagnosed preoperatively in one patient. The diagnosis was confirmed intraoperatively and at post mortem. In left ventricular long-axis planer view from parasternal location the aortic-mitral valve continuity could be demonstrated (Fig.-15 A,B). In short axis planer view obtained from parasternal position at the base of the heart, the aorta was right-sided and somewhat anterior to the pulmonary artery (Fig.-15-C). Correpative cine angiogram and anatomic post mortem specimen showed that the
A, Sector scan from a normal subject obtained from suprasternal location in the coronal plane, B, Images obtained by anterior tilting of the transducer, C, Similar view as A in a patient with d-transposition of the great arteries, and D, Pathologic heart specimen of a patient with d-transposition in identical plane as A. Note a close correlation of the echo image and anatomic heart specimen. The ascending aorta is seen in short axis above the right pulmonary artery (RPA) in the normal subject and from position A requires anterior and leftward transducer tilting motion to image the ascending aorta, B. In patient with d-transposition of the great arteries aorta is rightward than main pulmonary artery and from position C requires rightward and anterior tilting motion to image the ascending aorta, D.

FIGURE — 10.

FIGURE — 9.

A, Correlative left ventricular cineangiogram (antero-posterior view) and left ventricular long axis view sector scan from subxiphoid position in a patient with transposition of the great arteries and intact ventricular septum. Note the connection of the pulmonary artery to the left ventricle. Branching of the main pulmonary artery (MPA) into right and left pulmonary arteries identify this structure. The pulmonary artery arose entirely from the left ventricle and at its origin severe muscular stenosis was seen. The aorta overrode the ventricular septum (Fig. 16 A,B).

DISCUSSION

Our study showed that the arterio-ventricular connections can be prospectively imaged using a two-dimensional sector scan echocardiographic technique. Interarterial and arterio-ventricular relationships can be established using various planar views (2,3). In smaller children, due to a small intrapulmonary air interface, cardiac imaging can be done from almost all areas over the precordium.

This study and the reported experience shows that normally related great arteries, where the aorta is posterior and rightward connected to the left ventricle and the pulmonary artery anterior and leftward connected to the right ventricle, can be imaged from various locations. The recognition of normally related aorta in this study was based on its continuation into the aortic arch from where, at times, the cranial vessels were seen to arise. Left ventricular long axis planer-
A Sector scan obtained from subxiphoid position in four chamber plane, B, In left ventricular long axis plane and C, Correlative left ventricular cineangiogram in anteroposterior view in a patient with 1-loop 1-transposition and pulmonary atresia. Discordant atrioventricular connections are demonstrated (A); the ventricular septum is almost horizontal (B) and a moderately large ventricular septal defect (D) is present. Pulmonary artery (PA) is recognized by its branching pattern, atretic origin of the pulmonary artery is well noted on sector scan (arrow). Left ventricle is identified because of smooth septal surface and mitral valve attachments exclusively to non-septal left ventricular wall (A). On angiogram, the pulmonary artery is opacified through a Waterston-Cooley shunt.

FIGURE – 11.

A Sector scan left ventricular long axis planar view obtained from high left parasternal location in a patient with d-transposition of the great arteries. Note the origin of the main pulmonary artery (MPA) with its first branches originating from the left ventricle (LV).

B. Angiogram showing the same.

view obtained from parasternal location could be used to demonstrate connection of aorta to the left ventricle, however, the identity of the aorta often remained uncertain since aortic arch could not be visualized. High parasternal location allowed greater visualization of the ascending aorta but did not resolve the problem of aortic arch visualization. Subxiphoid location using long axis planar view was found to be most useful for imaging the aortic arch ascending aorta and its connection to the left ventricle.

Parasternal short axis planar views at the base of the heart helped to define not only anterior but also left and right interarterial relationships of the great arteries (2-7). In this view arterioventricular connections cannot be imaged which were best imaged from subxiphoid location. In large subjects, the use of subxiphoid location, at times poses difficulty, particularly in long chested individuals with narrow subcostal space. In these cases additional views from the precordium are mandatory.

The identification of normally related pulmonary artery was achieved from subxiphoid location using left ventricular short axis plane. The pulmonary artery to anterior right ventricular connection could be clearly established. From
FIGURE - 14.

A, B, Sector scan echocardiograms in coronal plane obtained from suprasternal location in a patient with 1-loop 1-transposition of the great arteries and intact ventricular septum. C, Left ventricular cineangiogram (anteroposterior view), and D, right ventricular cineangiogram of same patient as A.

Echocardiogram A, B shows left-sided ascending aorta (AAO) while main and right pulmonary arteries lie rightward. Leftward and anterior tilting of the transducer from position B is required to visualize the ascending AO as in A.

FIGURE - 13.

A, B, Sector scans in a patient with 1-transposition of the great arteries obtained from subxiphoid position in left ventricular long axis plane. The imaging is begun by first obtaining a four chamber image of the heart, the transducer is then tilted anteriorly and rightward as in B to image the pulmonary artery arising from the left ventricle above the ventricular septal defect (D). A leftward and anterior motion of the transducer is then required to image a leftward aorta connected to left-sided right ventricle (A).

parasternal location using short axis, planar views at the base of the heart, pulmonary artery and its branches could be imaged in all instances. Combining this view from two different locations, identification of the pulmonary artery could be established in all instances.

The suprasternal views demonstrated that the aorta and pulmonary artery could be identified by their characteristic anatomic relations. Furthermore, interarterial relationships could be accurately defined. Characteristic positions of the aorta and pulmonary artery were imaged not only for normally related great vessels but also for complete (d-TGA) transposition and corrected or 1-transposition of the great arteries. In 1-transposition of the great arteries, leftsided anterior aorta could be demonstrated in all subjects. We were able to use this location even in newborn subjects with relative ease. In our experience, this view provided excellent confirmation of the
imaging of great vessels from other positions.

The high left parasternal location was found to be useful in establishing both the interarterial and arterio-ventricular connections. The left parasternal view provides imaging of the cardiac structures which was correctly orientated anatomically vis-a-vis the body, the cardiac apex points to the left and inferiorly and in normally related great arteries, pulmonary artery is leftsided and anterior. The left atrium could be viewed from the superior to the inferior borders. Unlike the left ventricular long axis planar view obtained from standard parasternal location, the ascending aorta up to the arch could be seen without distortions. The transverse sulcus can at times be seen between the posterior wall of the aorta and anterior wall of the left atrium. In this view origin of the main pulmonary artery and its branches from the left ventricle was recognized in transposition of the great vessels.

Aortic displacement above the right ventricle, as noted in tetralogy of Fallot and double outlet right ventricle was demonstrated from subxiphid and left parasternal locations. This capability, we believe, makes the echo images invaluable in anatomically classifying the aortic or pulmonary arterial connection in double outlet right ventricle.

Subxiphoid (subcostal) and left parasternal locations demonstrated the feature of overriding aorta and infundibular stenosis with remarkable clarity. The extent of aortic overriding, position of the ventricular septal defect and degree of infundibular hypoplasia could be evaluated. Since the position of the ventricular septal defect vis-a-vis the great arteries could be determined, the anatomic diagnosis of this anomaly could be established by two dimensional echocardiography.

Complete transposition of the great arteries (d-TGA) was prospectively diagnosed in all of our patients. As has been reported, the anatomic recognition of pulmonary artery connected to left ventricle and aorta to right ventricle was possible in all instances (8). The associated defects, such as ventricular septal defect and subpulmonary stenosis were additionally visualized, when present. Although we established a prospective diagnosis of inverted or corrected 1-transposed great arteries in one of four cases, nonetheless diagnostic anatomic imaging was possible in all four patients. From subxiphoid location using four chamber and left ventricular long axis views, discordant inferior vena cava, right atrium, and rightsided left ventricular connections could be established. The left ventricle was recognized by the papillary muscle attach-
ments confined to the ventricular free wall and lack of papillary muscle attachments to the septum. The pulmonary artery arose from right-sided ventricle in a vertical fashion, recognized by its branching pattern. The right ventricle was seen as a triangular chamber with papillary attachments to both the free wall and intact ventricular septum, giving rise to left-sided aorta recognized by its continuation into the aortic arch. The ventricular septum and its oblique or horizontal alignment was remarkably well delineated. The position of the ventricular septal defect, when present, was well seen. We were able to obtain diagnostic imaging in a 17-year old patient with 1-transposed great arteries from subxiphoid location (Fig. ). Suprasternal position was confirmatory of 1-transposed great vessels in all subjects. Views from high left parasternal location also provided diagnostic anatomic features of 1-transposed great vessels.

Our experience with double outlet left ventricle is limited; however, the diagnosis seems to be possible preoperatively. It is concluded that two-dimensional sector scan echocardiography provides a precise means for the prospective diagnosis of the interarterial and arterio-ventricular relationships of the great arteries when all possible transducer locations are utilized to study these relationships.

References:


