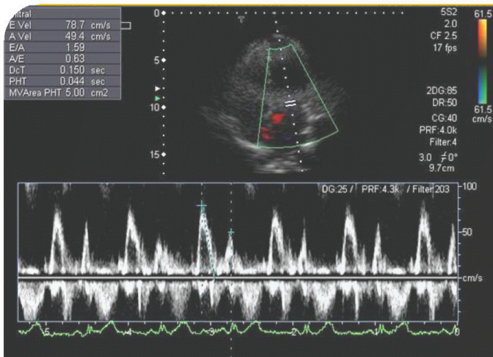

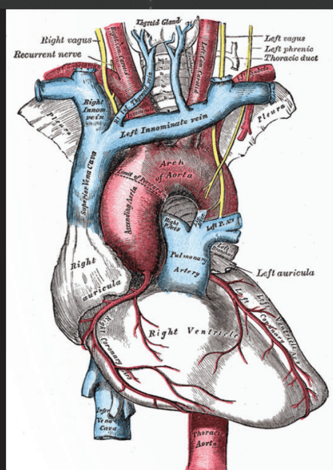

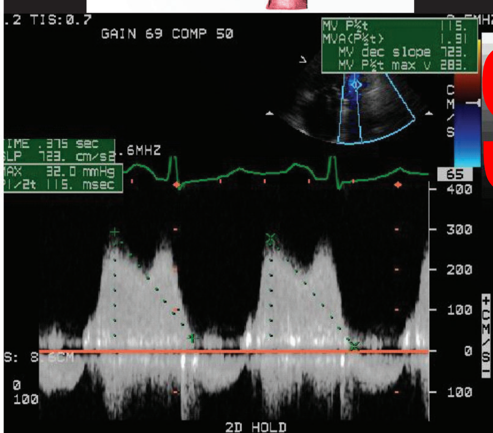

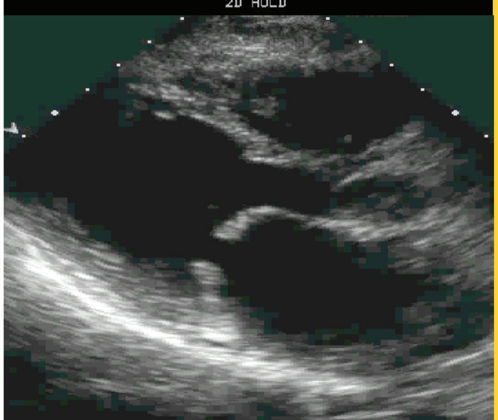

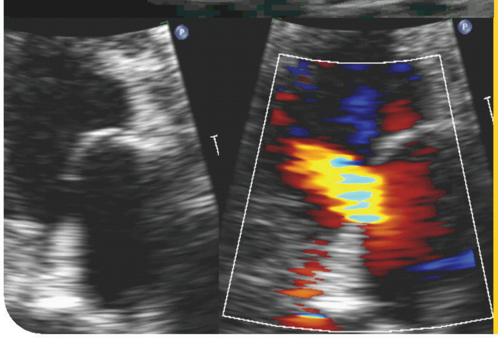

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GULFCOAST ULTRASOUND INSTITUTE INC

111 2nd Ave. NE Suite 800
St. Petersburg, FL 33701
Tel: (727)363-4500
Fax: (727)363-0811
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EDUCATIONAL NEEDS STATEMENT:
The planning committee has determined a need for this educational activity based on request from the medical community, expanded utilization of ultrasound, certification and lab accreditation requirements.

ENDURING MATERIALS COURSE OBJECTIVES:
Please see opening statements at each section.

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Author: Christie Jordan, BS, ACS, RDCS, RCS, RVS, RCIS, FASE
Cardiovascular Technology Program Director
Florida State College at Jacksonville, Jacksonville, Florida
No relevant financial relationships to disclose

Disclosure of Individuals in Control of Content
In addition to the faculty listed the following individuals are recognized by GUI as being in control of content of this program:

James Mateer, MD, RDMS (Medical Director-planner & QI Task Force)
Medical Director, Gulfcoast Ultrasound Institute
Milwaukee, WI
No relevant financial relationships to disclose

Charlotte Derr, MD, RDMS, FACEP (Co-Medical Director-planner & QI Task Force)
Assistant Professor of Emergency Medicine &
Fellowship Director of Emergency Medicine
Ultrasound Fellowship Program
University of South Florida Medical School
Tampa, FL
No relevant financial relationships to disclose

Andreas Dewitz, MD, RDMS (Member of Advisory Board & QI Task Force Subcommittee)
Associate Professor of Emergency Medicine Vice Chair of Ultrasound Education
Boston Medical Center
Boston, MA
No relevant financial relationships to disclose

Lori Green, BA, , RDMS, RDCS, RVT (Program Director-planner, Content Reviewer, QI Task Force)
Gulfcoast Ultrasound Institute, Inc.
St. Petersburg, FL
No relevant financial relationships to disclose

Trisha Reo, AAS, RDMS, RVT (Program Coordinator-planner, Content Reviewer, QI Task Force)
Gulfcoast Ultrasound Institute, Inc.
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ADULT ECHOCARDIOGRAPHY REGISTRY REVIEW WORKBOOK

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Section 1:

Anatomy and Physiology

Section 1: ANATOMY AND PHYSIOLOGY

Objectives

Upon completion of this module, you should be able to:

- Recognize and describe cardiac anatomy and physiology
- Identify the subdivisions of the ventricles
- State the normal pressures in all four cardiac chambers
- Compare electrical and mechanical systole
- Relate the filling phases of diastole to the cardiac cycle
- Compare timing of cardiac events to ECG
- Identify basic heart sounds

SEGMENTS OF THE HEART

Right Atrium

The heart receives deoxygenated blood from the venous system that returns this deoxygenated blood to the right side of the heart. This blood empties into the right atrium via the superior vena cava, inferior vena cava, and the coronary sinus.

The **superior and inferior vena cava** returns blood from the upper and lower regions of the body respectively. The **coronary sinus** drains deoxygenated blood from the heart. The superior and inferior vena cava enter the right atrium on the posterior aspect of the heart. The inferior vena cava has a rudimentary valve formed by a tissue fold called the Eustachian valve. The superior vena cava has no such valve. The coronary sinus enters the right atrium between the IVC and the tricuspid valve. It is guarded by a flap of tissue which forms a rudimentary valve called the Thebesian valve.

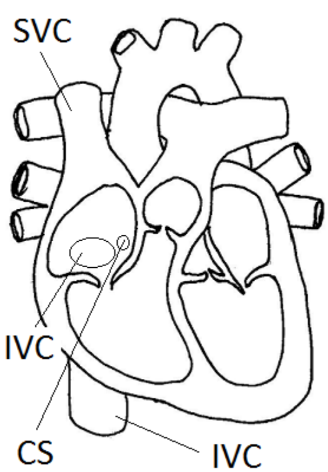


Image 1:1: Right-sided anatomy of the heart

The **right atrium** is located on the right, superior portion of the heart and forms the right lateral cardiac border. It is located behind and to the right of the right ventricle and lies mostly anterior to the left atrium. The right atrium can be divided into two sections: 1) a smooth section, and 2) a rough, trabeculated section. The smooth area of the right atrium is located posteriorly where the SVC and IVC enter the right atrium. There is an internal ridge that separates the smooth portion of the right atrium from the more trabeculated region called the crista terminalis, The trabeculated area of the right atrium that is usually composed of pectinate muscles houses the right atrial appendage which extends from the right atrium and drapes over the aortic root. The appendage is a triangular shaped pouchlike structure with a large orifice. Its purpose is to increase the size of the right atrium so it can hold a greater volume of blood.

From the right atrium blood passes through the **tricuspid valve.** The tricuspid valve is made up of three leaflets: 1) anterior, 2) septal (medial) and 3) posterior. This area is also called the **right ventricular inflow tract** as it directs blood into the right ventricle.

Right Ventricle

Once blood crosses the tricuspid valve it enters the **right ventricle**. The RV is typically the most anterior cardiac chamber and appears somewhat crescent shaped. The walls of the right ventricle are much thinner as compared to the walls of the left ventricle. The right ventricle walls are also lined with trabeculae carneae.

Both ventricles share the interventricular septum as their medial wall. The moderator band is a muscle commonly seen during an echocardiogram and is located in the right ventricle. The moderator band extends from the lower interventricular septum to the anterior wall and then joins the anterior papillary muscle.

The **right ventricular outflow tract** is also called the conus arteriosus or infundibulum. The crista supraventricularis, a thick muscle, separates the outflow tract from the inflow tract. The supraventricularis arches over the anterior leaflet of the tricuspid valve. The right ventricular outflow tract extends from the tricuspid annulus to the pulmonic valve. From the right ventricular outflow tract, blood crosses the **pulmonic valve** and enters into the pulmonary artery. The pulmonic valve is made up of three cusps: 1) anterior, 2) right and 3) left cusps.

The **pulmonary artery** returns deoxygenated blood to the lungs. The pulmonary artery begins at the pulmonic valve level where it bifurcates into the right and left pulmonary arteries a few centimeters from its origin. The pulmonary artery has thinner walls in adults, with pressures approximately one- sixth of the systemic circulation in the normal adult heart. Pulmonary capillary pressures at rest are only 7-10 mmHg as opposed to systemic capillary pressures which are 25 to 35 mmHg.

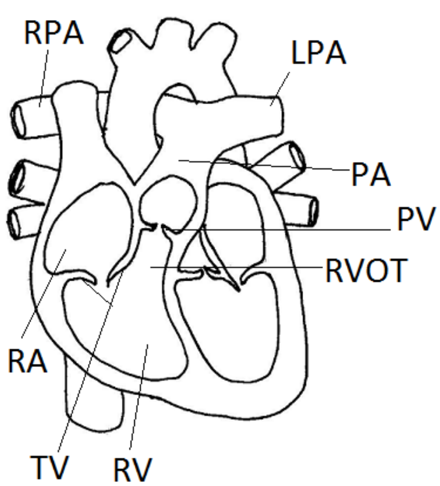


Image 1:2: Right-sided anatomy of the heart

Left Atrium

The **atria** are the receiving chambers for blood in the heart. The left atrium receives oxygenated blood from the pulmonary veins. There are normally four or five **pulmonary veins.** The left atrium also acts as a reservoir of the heart for ventricular systole, provides a significant amount of blood to the left ventricle during left atrial contraction, and is a conduit during left ventricular filling.

The **left atrium** lies superior and posterior to the other cardiac structures and is slightly smaller and thicker than the right atrium. The left atrial appendage is a projection of the left atrium from the anterolateral surface alongside the pulmonary artery. The left atrial appendage contains pectinate muscles while the rest of the left atrium does not. It is finger shaped and its opening has a narrow orifice. Like the right atrial appendage, its purpose is to increase the size of the left atrium to allow for an increase of volume blood flow. The left atrial appendage is an area of concern in patients with slow blood flow or in atrial fibrillation as this is a common area for a thrombus to form.

The **left ventricular inflow tract** is funnel shaped and formed by the mitral annulus, mitral leaflets, and chordae tendineae. The inflow tract directs blood from the left atrium to the left ventricle inferiorly and anteriorly.

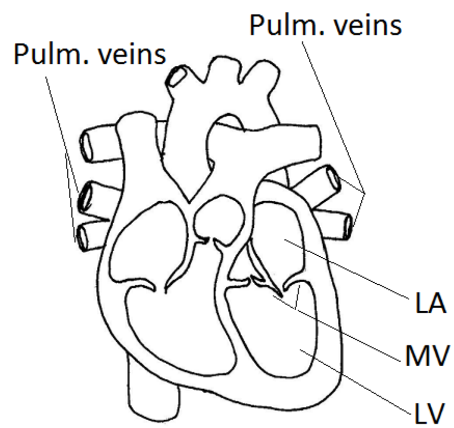


Image 1:3: Left-sided anatomy of the heart

Left Ventricle

Once blood crosses the mitral valve it enters the **left ventricle.** The left ventricle is the larger of the two ventricles, yet both the right and the left ventricle eject equal amounts of blood volume. It is the thickest chamber of the heart containing about 75% of the muscle mass of the entire heart and forms the apex of the heart. The left ventricle has an ellipsoid or conical shape and contains trabeculae carneae, although the left ventricular septal surface is smoother than the right ventricular septal surface which is heavily trabeculated.

The **left ventricular outflow tract** is surrounded by the inferior surface of the anteromedial mitral leaflet, the interventricular septum, and the left ventricular free wall. It ends at the level of the aortic annulus.

Blood passes from the left ventricle through the **aortic valve**. The aortic valve contains three cusps: 1) right coronary 2) left coronary and 3) non-coronary cusps. The names of the cusp are derived by where the coronaries arise off of the aorta in the sinus of Valsalva area.

After the blood crosses the aortic valve, it enters the aortic root where the oxygenated blood is distributed to the body. The initial area of the aorta is called the ascending aorta then arches around to form the arch of the aorta and into the descending aorta which runs down the body before bifurcating. There are three main branches at the arch area of the aorta 1) innominate or brachiocephalic which branches into the right subclavian and right common carotid arteries, 2) left common carotid and the 3) left subclavian branch.

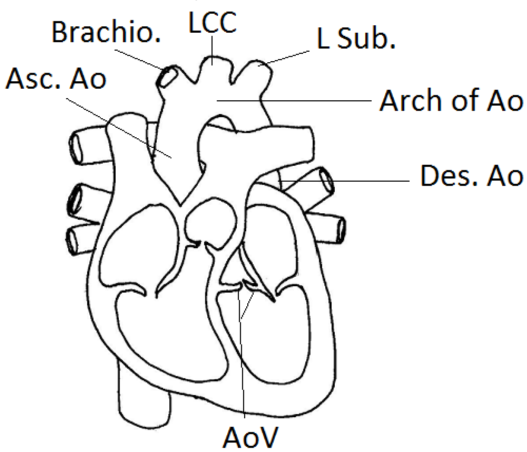


Image 1:4: Left sided anatomy of the heart

The set of heart valves that separate the atria from the ventricles are called the **atrioventricular valves**. These are the mitral and tricuspid valves. The tricuspid valve regulates flow between the right atrium and right ventricle. There are three leaflets composed of fibrous tissue covered by endocardium. The tricuspid orifice is larger than the mitral. The tricuspid leaflets are not equal in size. The anterior leaflet is the largest and extends from the infundibular region to the inferolateral right ventricular wall. The medial leaflet is the septal leaflet. It attaches to both the membranous and muscular interventricular septum. The posterior leaflet is usually the smallest and attaches to the tricuspid ring.

The **mitral leaflets** are similar in structure to the **tricuspid leaflets but** are thicker due to the higher pressures of the left ventricle. This is necessary to withstand the higher pressures in the left ventricle. The leaflets are attached at one end to the annulus fibrosis which completely circles the valve orifice. There are two mitral leaflets, the anterior leaflet is triangular and extends from the posteromedial left ventricle to the anterolateral left ventricular wall. This leaflet is continuous with the posterior aortic root wall.

The posterior mitral leaflet is smaller but longer leaflet and circles approximately two thirds of the mitral valve orifice. It attaches to the annulus fibrosus superiorly. The anterior and posterior mitral valve have two well defined indentions that separate the valve into three areas (scallops): A1 (anterior), A2 (middle), and A3 (posterior). The posterior mitral valve has the same indentions and corresponding names P1, P2 and P3.

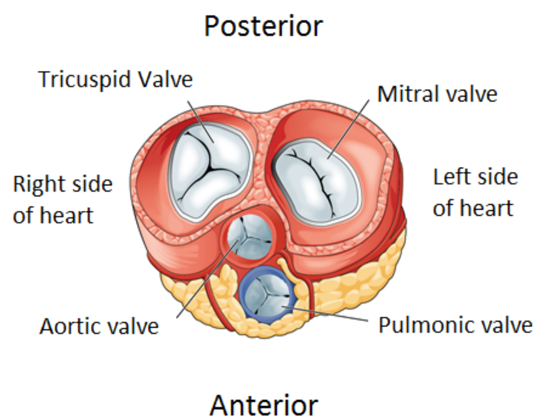


Image 1:5: Heart Valves

Chordae tendineae and papillary muscles are located in both the right and left ventricles.

There are usually three papillary muscles in the right ventricle, which have strong chords of fibrous tissue called **chordae tendineae** arising from their tips. The chordae attach the valve leaflets to the papillary muscles. The **papillary muscles** pull the valve leaflets together and downward during isovolumetric ventricular contractions. The tricuspid valve is usually more apically located than the mitral valve and has chordal attachments to the interventricular septum

ANATOMY AND PHYSIOLOGY QUIZ

1. Rapid depolarization correlates with this ECG waveform:
   a. S wave
   b. T wave
   c. P wave
   d. QRS complex

2. What phase of the cardiac cycle demonstrates four valve closures, ventricular contraction and increasing pressure?
   a. Isovolumetric relaxation
   b. Ventricular systole
   c. Isovolumetric contraction
   d. Ventricular diastole

3. The area of the heart where the coronary sulcus meets the interventricular sulcus is called the:
   a. apex
   b. crux
   c. coronary sinus
   d. AV groove

4. The inner surface of the heart wall is the:
   a. myocardium
   b. epicardium
   c. visceral layer
   d. endocardium

5. Which of the heart wall layers is responsible for the heart's ability to contract?
   a. parietal pericardium
   b. myocardium
   c. epicardium
   d. endocardium

6. The left ventricular outflow tract ends at the level of the:
   a. aortic annulus
   b. left ventricle
   c. papillary muscles
   d. chordae tendineae

7. The amount of blood that is pumped out of the ventricles with each beat is defined as:
   a. Cardiac Output
   b. Stroke volume
   c. End diastolic volume
   d. End systolic volume

8. What structure in the right ventricle extends from the lower interventricular septum to the anterior wall where it joins the papillary muscle?
   a. chordae tendineae
   b. tricuspid valve
   c. moderator band
   d. trabeculations

9. The right ventricular outflow tract is also referred to as the:
   a. conus arteriosus
   b. crista supraventricularis
   c. infundibulum
   d. b and d
   e. a and c

10. What are the normal number of pulmonary veins?
    a. 6 to 7
    b. 2
    c. 3
    d. 4 to 5

11. Where are the pectinate muscles located in left atrium?
    a. whole left atrium
    b. pulmonary veins
    c. posterior left atrial wall
    d. left atrial appendage

12. The cardiac structure that forms the right lateral cardiac border is the:
    a. left ventricle
    b. right ventricle
    c. right atrium
    d. left atrium

Section 2:

Technique

Section 2: TECHNIQUE

Objectives

Upon completion of this module, you should be able to:

- Optimize equipment controls
- Recognize commonly seen technical artifacts
- Integrate standardized imaging views to perform a two-dimensional cardiac ultrasound exam
- Identify regional wall segments
- Recognize normal M-mode patterns
- Utilize optimal scanning views and appropriate interrogation angles for performing Color Doppler Imaging and Spectral Doppler Imaging

TECHNIQUE

USE OF EQUIPMENT CONTROLS

The **depth** control affects several parameters relating to the ultrasound image. For adult echocardiography it is recommended by the ASE to begin with the "deep" view at around 20 – 24 cm and then change to an imaging depth of approximately 15 to 16 cm for a parasternal long axis (PLAX). The deep view allows the sonographer to see deeper behind the heart to differentiate or diagnose a pericardial or pleural effusion. The optimum depth is directly dependent on the size of the patient and transducer position.

Images should be taken at the shallowest depth as this allows for an optimal frame rate. Increasing the depth results in a decrease in the pulse repetition frequency since it will take longer for the pulse to travel back and forth from the transducer. The effect on frame rate should be minimal. When performing pulsed wave Doppler, the PRF is important in that it determines the Nyquist Limit. Pay close attention to the depth if you are performing PW Doppler and the waveform is aliasing.

Image 2:1: Sample of Depth of Image

The **amplification** of the sound is controlled by the overall gain and TGC (time gain compensation). If the entire image is too bright, consider decreasing the power. If the entire image is too dark, consider increasing the overall gain (amplification)

The TGC controls can also selectively adjust the amplification or brightness of the image in specific areas or regions based upon depth. This is typically controlled by the use of slide pods or toggle switches associated with these regions of the ultrasound image.

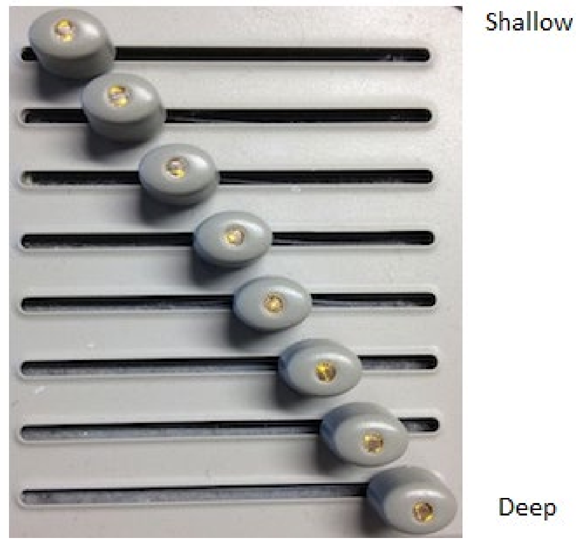


Image 2:2: Sample of TGC keys on ultrasound system

Near and far field gain controls may also be used to selectively adjust those areas respectively. Rotary knobs may also be used for these functions and for the overall gain.

Transmit power is the voltage which is applied to the transducer itself. This controls the intensity of the sound beam. Transmit power should be kept at lower settings following the ***"ALARA"*** principle; ***As Low As Reasonably Achievable*** since it may be associated with some biological effects.

The **focal zone** is the region where the best resolution can be obtained. Most systems allow the operator to adjust or move the focal zones on a real time image. Altering the focal zone allows for the beam to be collimated for a specific area of the image. Beyond the focal zone the beam diverges and therefore it should be set at or just below the main structure or area of interest in your field of view. Setting the focal zone too anterior to the structure of interest places it in the divergent portion of the beam which reduces its effectiveness. For example, from an apical four chamber view when interrogating the apex, it should be set in the near field and when interrogating the mitral valve, the focus should be set more in the far field. New systems typically **have an automatic focus.**

Sector Width of the transducer is also important. Widening the sector width will cause a decrease in the frame rate. Always keep the sector width as narrow as possible to maintain higher frame rates.

Dynamic range also known as Compression**,** in gray scale imaging is the smallest to largest group of intensities which produce a shade of gray on the monitor. Most equipment has a larger dynamic range than the human eye can perceive. The greater the dynamic range, the more potential gray scale shades. If the dynamic range is reduced, the shades of gray are decreased and the image contrast increases which may improve edge detection of the endocardium and make the chambers look more cystic.

Pre-processing allows for the altering or changing of the signal before the image is frozen while post processing alters the signal afterwards. Pre and Post processing will sometimes help to enhance an image that is technically limited.

The **Doppler angle** in cardiac, as well as in all ultrasound applications, is optimum when it is at zero degrees to flow or parallel to flow. The incident angle should be no greater than 20^0 to flow when performing Spectral Doppler. Doppler angles greater than 20^0 result in large calculation errors. The best rule to consider when performing any type of Doppler exam is, the lower the angle of incidence to flow, the better the returning or reflected signal. Angle correction controls allow for the exact measurement of the angle of incidence relative to the direction of flow. Angle correction controls are typically not utilized for cardiac Doppler since the Doppler views used are typically between 0^o and 20^o and the difference in the cosine value of 0^o to 20^o is very minimal. *See the following Cosine table.*

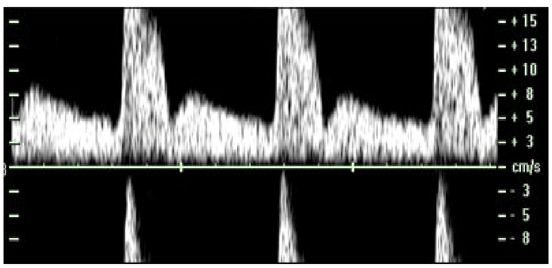
Cosines for Various Angles

Angle A (Degrees)	cos A
0	1.00
5	0.996
10	0.98
15	0.97
20	0.94
25	0.91
30	0.87
35	0.82
40	0.77
45	0.71
50	0.64
55	0.57
60	0.50
65	0.42
70	0.34
75	0.26
80	0.17
85	0.09
90	0.00

Table 2:1: Cosine of angles

The **sample volume or sample gate** is used for pulsed Doppler modalities. The sample gate or range gate is site specific and gives flow information specific to the area or the vessel that it is placed. The term used to describe this function is sometimes referred to as range resolution. The gate length may be increased or decreased to allow for a larger or smaller sampling area. The range of the sample gate is usually 1-20 mm. A longer gate usually improves the signal to noise ratio. Sample gate or sample volume width is always equal to the beam width at the site of the sample volume.

Zero shift or baseline controls allow for the baseline to be moved up or down depending on the Doppler signal, direction of the signal, and signal velocity. If a Doppler signal is cut off or aliases, the baseline can be adjusted to try to include the entire spectral signal. When the Doppler signal continues to cut off after the zero baseline is adjusted as much as possible, the pulse repetition frequency or PRF can be increased. When this fails to include the entire Doppler signal on the spectral tracing, continuous wave Doppler should be implemented to resolve the aliased signal.



Baseline needs to be shifted down to avoid aliasing

Image 2:3: Baseline shifting

Pulse repetition frequency or PRF may be referred to as velocity range or scale. The pulse repetition frequency is the number of pulses per second or other specified unit of time. When the pulse repetition frequency is increased, higher velocity signals can be detected. When the pulse repetition frequency is decreased the velocity range is decreased improving sensitivity to slower flow. PRF is also dependent upon depth of the image. Nyquist limit is equal to ½ of the PRF.

Spectral analysis is the breaking up of the parts of a complex wave. These parts are spread out in the order of their frequencies. With the use of FFT or Fast Fourier Transformer, the Doppler spectrum is derived from the various frequencies of the returning signals. This technique can demonstrate frequency, amplitude, and spectral broadening that results from a wide range of Doppler shifted frequencies.

On the spectral display, time is displayed on the horizontal axis corresponding to changes in the cardiac cycle and frequency shifts towards (+) or away (-) from the transducer are displayed above or below the zero baseline respectively.

Although the system is detecting frequency shifts, most newer systems display the calculated velocities on the vertical axis assuming the incident angle was 0° to 20° and thus using a cosine value of one.

The brightness of the gray scale spectrum analysis represents the number of events at that Doppler shifted frequency. Spectral broadening refers to the range of frequencies at any moment in time and is relative to the vertical axis of the spectrum analysis. A thicker waveform would indicate spectral broadening.

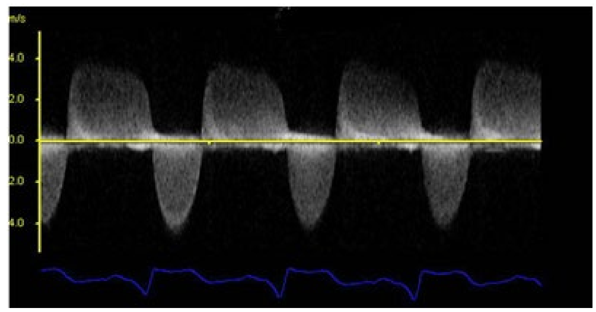


Image 2:4: Spectral display of forward and backward flow

The **resonant frequency** of a transducer is its operating frequency. The propagation speed of the transducer material and the thickness of the transducer element determine its operating frequency. Most adult echocardiography is performed with transducer frequencies of 2.0 to 5.0 MHz. The higher the frequency of the transducer the better the resolution, but penetration is decreased. Broad bandwidth or multi- frequency transducers allow a variety of both selectable imaging and Doppler frequencies for optimal resolution and penetration.

With the invention of tissue harmonic imaging, the sonographer can now obtain higher resolution images at lower operating frequencies. The soundwave is sent in at a lower frequency to increase penetration and harmonizes off of the tissue allowing the returning frequency to be doubled. All machines now use harmonic frequency and the frequency is typically displayed as 1.7/3.3 MHz. This means that the frequency going into the body is at 1.7 MHz for increased penetration and comes back to the transducer at 3.3 MHz which is a higher resolution image.

There are several types of transducers utilized in ultrasound imaging. A commonly used transducer in adult echocardiography is a **phased array** transducer. This type of transducer has several elements in the transducer head. Voltage pulses are applied to all of the elements, but at small time differences which allows the beam to be shaped and steered. Phased array transducers are quiet, lightweight and have more flexibility of beam angulation, field of depth, and sector width.

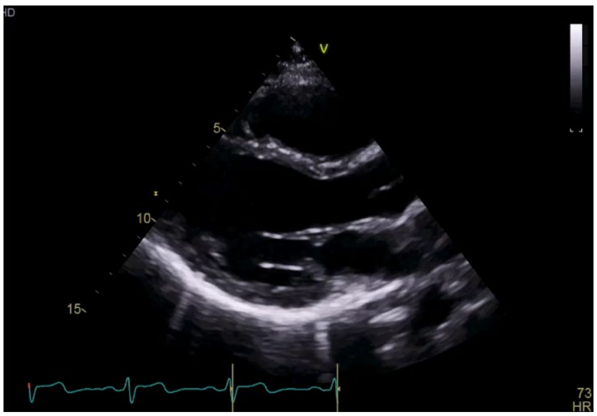


Image 2:5: Sample of Sector Shaped Image

Matrix array transducers are electronically steered and have on average 2,000 to 3,000 elements. Phasing the large number of elements arranged in a matrix enables the acquisition of several scan planes simultaneously. For instance, both the parasternal long and short-axis or apical 4 chamber and 2 chamber 2D scan planes of the heart can be shown. In addition, it allows an individual scan plane to be swept or rotated through a 3-Dimensional volume of tissue.

Play Video V2:1: Matrix Array Beam animation

In addition to their multiplane view and 3D applications, most of the matrix array transducers have some 4D capabilities too which offer both current and future advantages for cardiac applications.

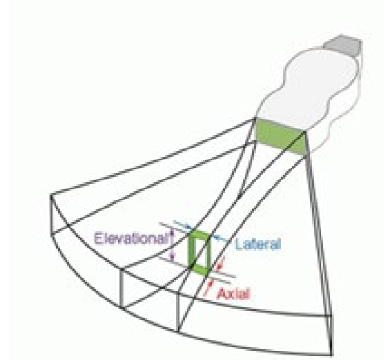


Image 2:6: Sample of Matrix Array Transducer

RECOGNITION OF TECHNICAL ARTIFACTS

Artifacts in ultrasound are anything on the picture or display which does not belong to the structures being imaged and are not real.

Reverberation artifacts are high-amplitude, linear artifacts that result from two strong specular reflectors. These create a back-and-forth reflection of the ultrasound before it can return to the transducer creating a redundant malposition series of reflectors going in different directions from the main sound beam. This results in reflectors being placed on areas in the wrong location on the image. Multiple reflectors may be noted in different locations on the image.

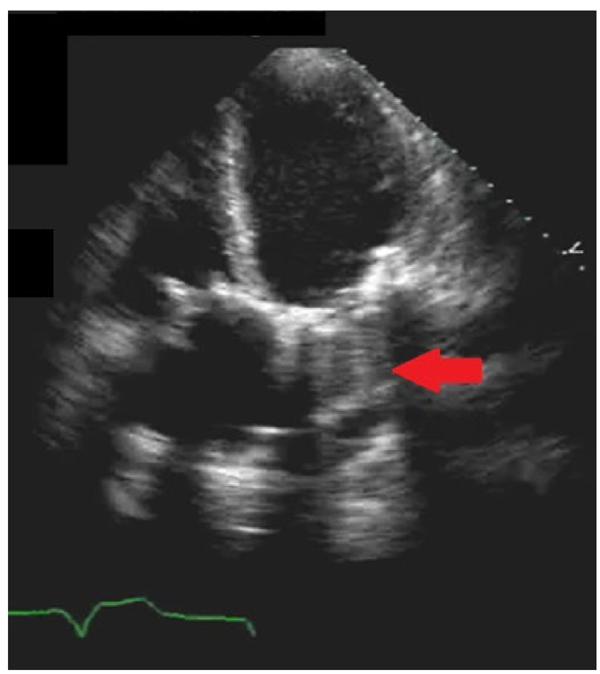


Image 2:7: Reverberation Artifact from Mechanical Valve

Mirror images are often seen in cardiac imaging. Mirror images are seen when objects which are on one side of a strong reflector are also noted on the other side.

Side lobe artifacts are caused by the acoustic energy emitted by the transducer going in different directions from the main sound beam. This results in reflectors being placed on areas in the wrong location on the image. Multiple reflectors may be noted in different locations on the image.

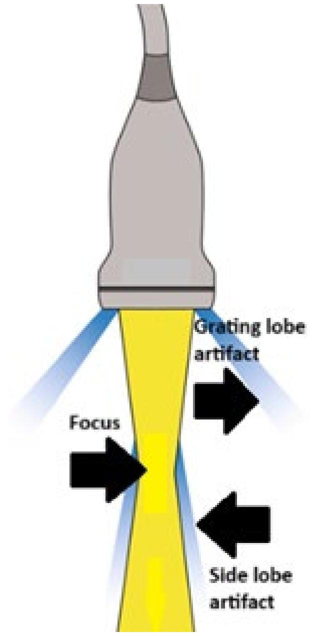


Image 2:8 Side lobe artifact

Acoustic shadowing may occur as a result of the sound beam encountering dense objects such as bone or calcifications in the body. These structures impede the sound causing attenuation that results in an acoustic shadow or drop out effect.

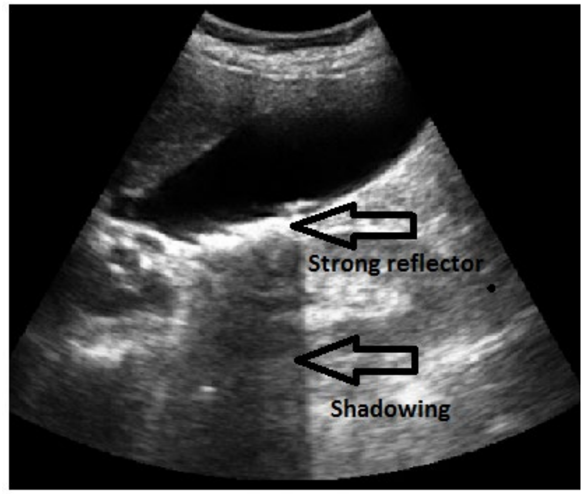


Image 2:9: Shadowing from a strong reflector

Aliasing of the color signal occurs when the flow exceeds the Nyquist limit. Color flow Doppler aliasing occurs at lower velocities than conventional pulsed Doppler. Color Doppler aliasing is seen as one-color folding over into the opposite color. Once aliasing patterns are recognized however, this artifact does not pose a significant problem and can help with the detection of certain cardiac anomalies. Aliasing alone is not a diagnostic criterion to indicate pathology.

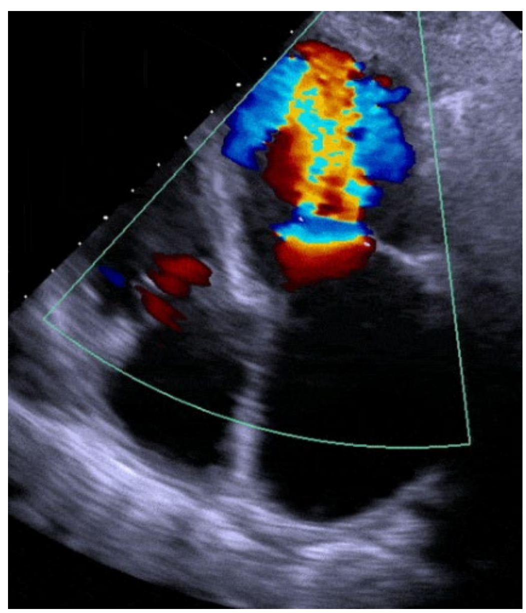


Image 2:10: Aliasing of Color Signal with Mitral Stenosis

Wall motion of the heart can produce signals that are stronger than the signals being produced by the red blood cells, resulting in an artifact. Some of this can be eliminated with the use of reject and wall filters. Color flow imaging may not be as sensitive as conventional pulsed Doppler causing weak signals to be missed. When the Color gain is increased color noise can be created. Color noise appears as a speckling of color that can occur inside or outside of the structure being evaluated.

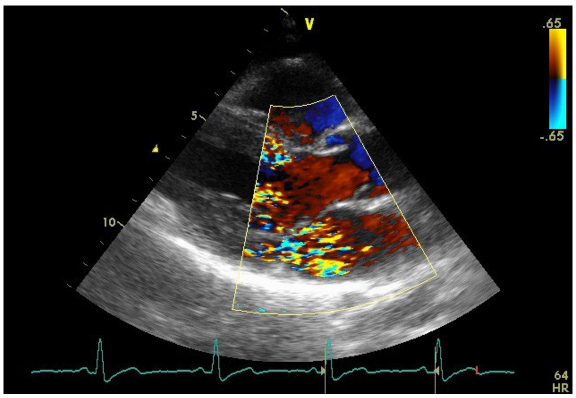


Image 2:11: Color Speckling or Noise

Temporal ambiguity may result when lower frame rates are utilized for color flow Doppler. Lower frame rates are used with color due to the increased time it takes to create a color flow image. The size of the sector can impact frame rate. A narrower, color sector or ROI (region of interest) box allows for faster frame rates and a wider sector or color ROI box decreases or slows frame rate.

TWO-DIMENSIONAL STUDY

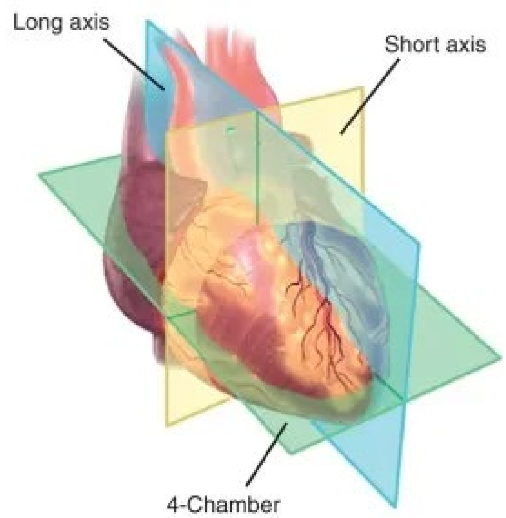


Image 2:12: Echo Imaging Planes

The basic echocardiogram usually includes four scan planes. The parasternal position is usually the starting point. The transducer is optimally placed in the 3rd to 4th intercostal rib space and as close to the sternum as possible. This allows for the most accuracy when obtaining measurements. Variations of this position can be used to show pertinent cardiac anatomy and abnormalities.

In the **parasternal** position the heart can be viewed in numerous scan planes. The standard planes are the parasternal long and short axis views. The **long axis** view images the heart in a lengthwise or longitudinal plane. With the Parasternal Long Axis view (PLAX), the transducer is placed in the 4LIS with the indicator marker pointing towards the right shoulder. The structures usually noted in this view are the right, ventricular outflow tract which is the most anterior structure. Below the right ventricle lies the interventricular septum (anteroseptal wall), left ventricle, and the inferolateral wall.

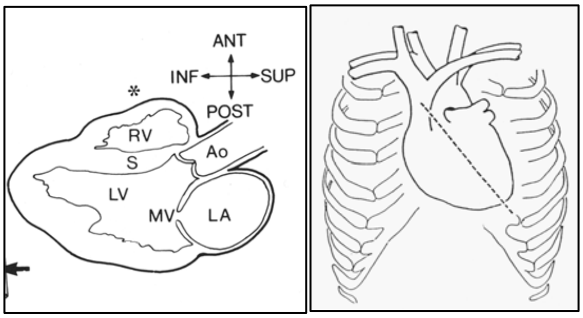


Image 2:13: Parasternal Long Axis view diagram and orientation

If the transducer is moved inferiorly and laterally the apex may be seen. Below the RVOT is the aortic root and valve, left atrium and mitral valve. The descending aorta is seen in cross- section posterior to the left atrium and is seen as a circle. The coronary sinus may also be noted between the left atrium and mitral valve, within the pericardium.

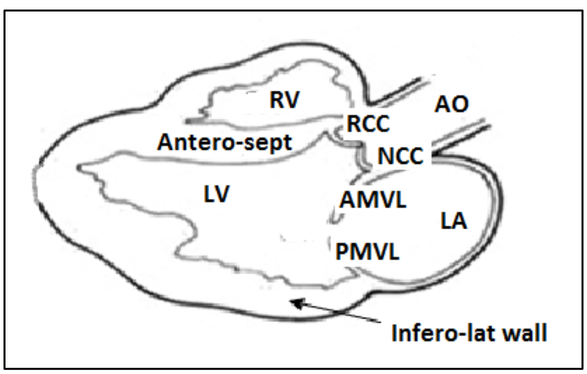


Image 2:14: Anatomy of the PLAX view

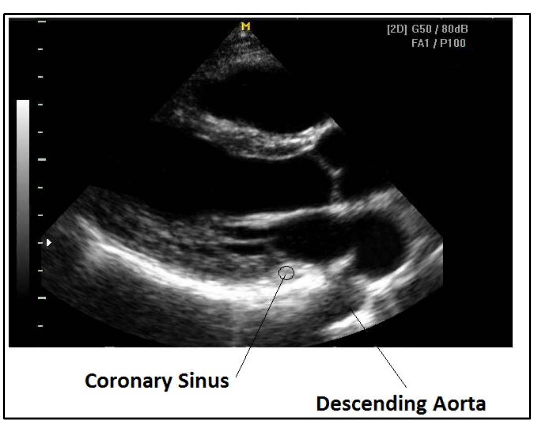


Image 2:15 Anatomy of CS and Desc. Aorta

Play Video V2:2: Parasternal Long Axis and PLAX RV Inflow View

The next image obtained is the right ventricular inflow tract view (RVIT). The transducer is tilted slightly inferiorly (towards the patient's right hip) In this view you can see the right atrium, tricuspid valve, the ostium of the coronary sinus and IVC in some patients and the right ventricle. Typically, from this view you will see the anterior tricuspid valve leaflet and the septal tricuspid valve leaflet (if the septum and LV still appear in view) then the anterior and septal TV leaflets are seen. By aiming more posteriorly, the posterior tricuspid leaflet is seen instead of the septal leaflet.

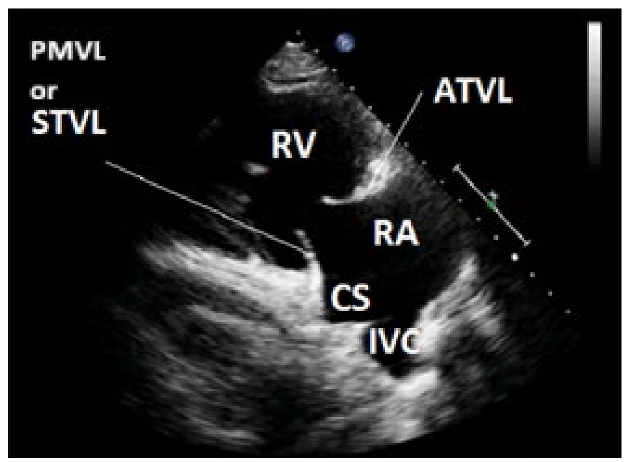


Image 2:16: Anatomy of RVIT view

The Right Ventricle Outflow View (RVOT) is the next view to image. This view is obtained by angling the scan plane from the PLAX superiorly with a slight counterclockwise rotation to visualize the RVOT, PV and proximal pulmonary artery.

Play Video V2:3: PLAX RV Outflow View

Section 10:

Congenital Heart Disease In the Adult

Section 10: CONGENITAL HT DISEASE IN THE ADULT

Objectives

Upon completion of this module, you should be able to:

- Identify various types of congenital heart diseases found in adults
- Recognize the types of aortic stenosis
- State the etiologies of congenital heart disease
- Recognize the types of atrial and septal defects and their associated findings
- Apply the echocardiographic methods to evaluate cardiac shunts, valvular stenosis, and related regurgitant lesions
- Relate knowledge of normal cardiac hemodynamics to the abnormal effects of the various congenital heart diseases

BASIC EMBRYOLOGY

PRIMITIVE HEART TUBE

Early in embryonic cardiovascular development, a pair of heart tubes is formed from the primitive vascular system of the fetus. A section of the main vascular channel specializes and develops contractile properties within its vessel walls. These heart tubes lie parallel to each other and in close proximity at the cephalic end of the fetus. Eventually, the pair of heart tubes fuses into a single endocrinal tube that retains its contractile properties. The wall of the primitive heart tube consists of an external myocardial mantle 1 to 2 cell layers thick. In addition, the wall also consists of a single layer of endothelial cells and is separated from the myocardial mantle by a basically structure less, cellular, third layer called cardiac jelly. The primordial heart is developed from the heart tube. The heart tube can be separated into sections by their responsible development. The sections consist of:

1) Truncus arteriosus
2) Bulbus Cordis
3) Primitive Ventricle
4) Primitive Atrium
5) Sinus Venosus

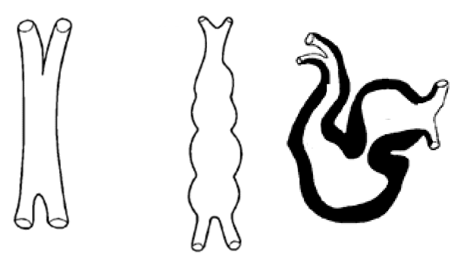


Image 10:1: Primitive heart tube

The Bulbus cordis is divided into three sections: primitive right ventricle, ventricular outflow tract and truncus arteriosus. The septum development divides the truncus arteriosus and bulbus cordis which becomes the aorta and pulmonary artery. The Primitive Ventricle becomes the left ventricle, the primitive atrium becomes the left and right atrium and the sinus venosus will become the IVC, SVC, coronary sinus and posterior wall for the atria.

Cardiac loop

At approximately day 23 in embryonic development, the growing heart tube is referred to as the **bulboventricular tube,** and bends to the right and anteriorly to form a sigmoid shape. This process is called **cardiac looping** and is believed to be a fundamental and intrinsic property of the myocardium, rather than a space saving adaptation in the rapidly growing fetus. At the same time and considered a part of cardiac looping, the aortic arches are developed in the cephalic portion of the bulboventricular tube. At the caudal half of the heart tube, tissue expansion forms the early version of the adult ventricle. The primitive atria lie extra pericardial and caudal to the ventricles. Looping adds a certain amount of torsion to the developing heart which is responsible in part for the spiral disposition of the interventricular septum.

Aortic arches

In the fetus, simultaneous with cardiac looping is a process which results in the formation of six arches which make up the aortic arch system.

These arches develop from the aortic sac portion of the primitive heart tube and come and go with the ever- changing structures of the fetal heart. These six arches are functional although only arches number three, four, and six persist in the adult heart and become the following:

- Number three becomes the carotid arteries
- Number four forms the aortic arch
- Number six forms the pulmonary arteries and ductus arteriosus

Septation

Cardiac septation is the process dividing the rapidly growing and expanding portions of the heart tube into the four distinct chambers of the adult heart. Septation takes place more or less simultaneously with the development of the primitive ventricles, conus cordis, and truncus arteriosus.

Septation is the result of three mechanisms

1. **Passive septation** is the fusing together of the growing and expanding wall segments of the primitive heart chambers.

2. **Active septation** is the growth of a layer of cardiac mesenchyme called endocardial cushion tissue. This tissue is derived from the earlier cardiac jelly and grows new cells which fuse together to form parts of the septa in the heart.

3. The third process of septation is a **combination** of the first two methods. This method starts out as passive septation only to have growth completed by endocardial tissue along borders which grow and fuse together.

Partitioning of the embryonic heart is completed by the formation of seven septa, three of which are passively formed, three of which are actively formed, and one that is formed by a combination of both methods.

Valve formation

Valve formation in the embryonic heart is accomplished primarily by infolding and growth of the muscular ventricular wall which is facilitated by the process of **diverticulation**. The remaining growth is completed by the expansion and fusion of endocardial tissue. Early in the development of the atrioventricular valves, they are thick and fleshy only to transform into the thin, fibrous cusps seen in the adult.

COMPARISON OF FETAL AND POSTNATAL CIRCULATION

Upon completion of the embryonic development of the fetal heart, there are still a few differences that stand out from the postnatal state. Fetal cardiovascular needs are met by certain modifications that differentiate it from post-natal circulation. In the fetus, highly oxygenated blood returns from the placenta in the umbilical vein. Half of this blood passes through the fetal liver and the other half through the inferior vena cava via the **ductus venosus**. After a short course through the inferior vena cava, the saturated blood enters the right atrium. The saturated blood is mixed with desaturated blood returning from the superior vena cava.

The design of the right atrium directs the majority of saturated blood through the **foramen ovale** into the fetal left atrium. A small percentage of the mixed, but still highly-saturated blood, crosses the tricuspid valve, passes into the right ventricle, and is ejected out of the pulmonary artery. Some of the blood goes to the lungs, but the majority is shunted to the descending aorta through the **ductus arteriosus**.

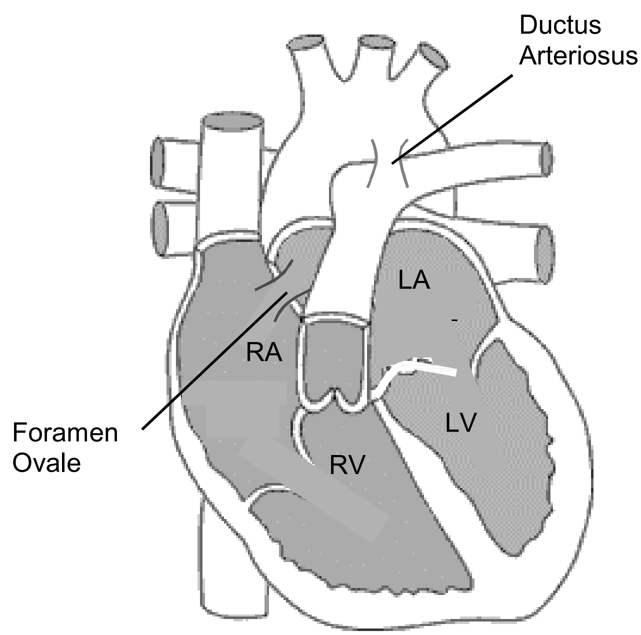


Image 10:2: Cardiac fetal circulation

Shortly after birth the three shunts present in the fetal circulation cease to function. The ductus venosus constricts so all blood entering the liver must pass through the hepatic sinusoids. Occlusion of the placental circulation causes an immediate fall in blood pressure in the inferior vena cava and right atrium. Aeration of the lungs with the first breath causes a dramatic fall in pulmonary vascular resistance and an increase in pulmonary blood flow. The second shunt closes when increased pulmonary blood flow causes left atrial pressure to rise, closing the flap of the foramen ovale. The third shunt is the ductus arteriosus which constricts at birth with the presence of bradykinins. Bradykinins is a substance released in the lungs with their initial inflation.

It is important to remember that the change from fetal to adult circulation is a gradual occurrence that often occurs over weeks. During the transitional stage there is often some flow through the three fetal circulatory structures.

During the complex stage of embryologic development of the heart, congenital lesions may develop which may or may not constitute a serious threat to life. Approximately 8 per 1000 live births result in some kind of congenital cardiac abnormality. Of these births, about 1/3 or 2.6 per 1000 births manifest critical heart disease.

The most common explanation as to why congenital defects occur is that they are due to a combination of genetics and environmental interactions. Risk to the fetus is increased with a genetic predisposition, as well as exposure to an environmental teratogen during a critical or vulnerable period of development. Children of parents with certain types of congenital defects are at a higher risk, with an approximately 8.8% chance of developing significant cardiac lesions.

ABNORMALITIES OF SEPTATION

Congenital cardiac defects in the newborn can be classified into four categories:

1. Abnormalities of septation
2. Abnormal vasculature
3. Valvular abnormalities
4. Persistence of fetal circulation

A ventricular septal defect is the most common abnormality, followed by pulmonary stenosis, patent ductus arteriosus, atrial septal defect (secundum type), and aortic stenosis

Abnormalities of Septation (Atrial)

There are three types of atrial septal defects which are classified as to their location, they are: **venosus**, **secundum**, and **primum** defects. The atrial septum is divided into three regions based on embryologic derivation of the area.

A **sinus venosus atrial septal defect** is located at the most superior portion of the atrial septum. It is bordered by the posterior wall of atria, the right atrial appendage, and the limbus of the fossa ovale. It lies in close proximity to the right sided pulmonary veins and superior vena cava. This defect is sometimes associated with anomalous pulmonary venous return.

A **secundum or fossa ovalis defect** is the most common atrial septal defect occurring in approximately 75% of babies. It is located inferior and anterior to the sinus venosus region. The secundum region is the thinnest portion of the atrial septum and is composed of the flap which covers the foramen ovale.

Primum or atrioventricular defect is bordered by the atrioventricular valves, posterior atrial wall, and the fossa ovalis and occurs approximately 15% of the time. Primum defects are usually associated with a cleft mitral valve.

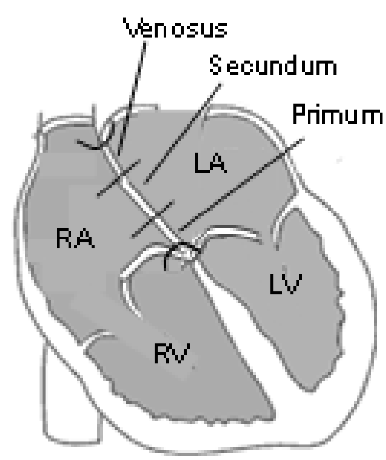


Image 10:3: Location of atrial septal defects

Abnormalities of Septation (Ventricular)

The other types of septal defects are termed ventricular septal defects. There are four types of ventricular septal defects:

1) Perimembranous
2) Trabecular (muscular)
3) Inlet (atrioventricular canal)
4) Outlet (supracristal, subpulmonic)
5) Malalignment

Perimembranous VSD's are the most common adult defect and may be associated with aortic regurgitation. This type of VSD is best seen in the PLAX, PSAX AoV level and the A5C view. In the PSAX view at the aortic level, the VSD is seen in the 10 – 12 o'clock position.

Trabecular VSD's are the second most common adult defect and may have multiple lesions. Trabecular VSD's can be seen in the PSAX view at the LV level, and the A4C and A5C view.

Inlet VSD's are best seen in the PSAX LV level and A4C view. This needs to be differentiated from a perimembranous VSD as the two defects are close in proximity.

Outlet VSD's are commonly seen in the Asian population and have a strong association with aortic regurgitation. In the PSAX view at the aortic level, the VSD is seen in the 12 - 3 o'clock position.

A **Malalignment** VSD are those types of VSD's seen in Tetralogy of Fallot or Truncus Arteriosus.

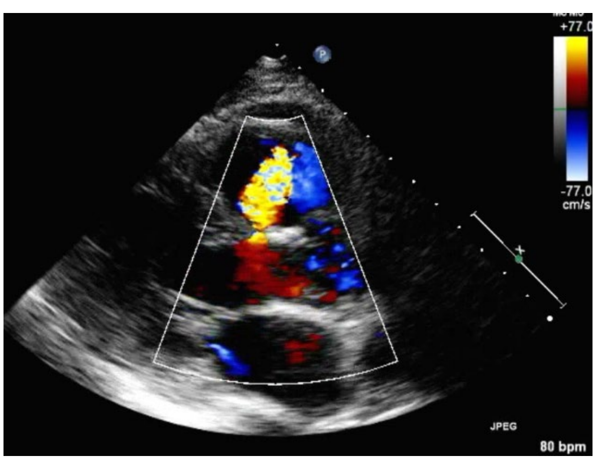


Image 10:4: Perimembranous VSD seen in PLAX view

A right ventricular systolic pressure can be obtained by the velocity of the ventricular septal defect.

RVSP (mmHg) = Systolic BP - $4V^{2\ (\text{VSD Peak Vel})}$

Abnormal Vasculature and Resulting Lesions

In addition to defects in cardiac septation, the embryologic heart may develop abnormal vasculature and communication between cardiac structures. Some of the more well-known include the following:

- Patent Ductus Arteriosus
- Anomalous Pulmonary Venous Return
- Coarctation of the Aorta
- Transposition of the Great Arteries
- Truncus Arteriosus

Anomalous pulmonary venous return involves the effective attachment of one or more of the pulmonary veins to some other structure other than the left atrium.

Coarctation of the aorta is an abnormal narrowing of this vessel usually just beyond the left subclavian artery.

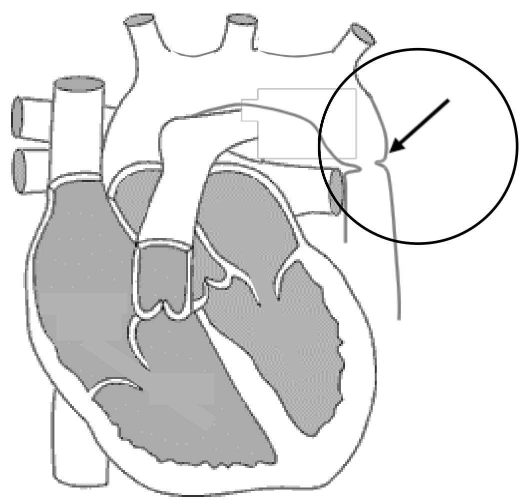


Image 10:5: Coarctation of the Aorta

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