Recommendations for the Adult Cardiac Sonographer Performing Echocardiography to Screen for Critical Congenital Heart Disease in the Newborn: From the American Society of Echocardiography

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BACKGROUND/NEED FOR DOCUMENT

Congenital malformations are the leading cause of infant mortality in developed countries, with critical congenital heart disease (C-CHD) being the major contributor to death and morbidity despite the development of specialized pediatric cardiac centers.[1,2] C-CHD is defined as congenital heart disease requiring surgery or catheter intervention in the first year of life and constitutes ~25% of CHD.[3] Although CHD is the most common form of congenital malformation and occurs in 9 of every 1,000 live births,[4] it is not always identified early and referred to a pediatric cardiologist. There is, therefore, a need for all cardiac sonographers, regardless of their pediatric experience, to be able to detect CHD and recognize those cases that are critical in nature.

Despite advances in antenatal screening and fetal echocardiography, prenatal detection of CHD remains variable by geographic location and type of CHD lesion, with a recent report from the United States (US) estimating a detection rate of only 42% in 2012.[5-7]

ABBREVIATIONS

AV = Atrioventricular
AoV = Aortic valve
CHD = Congenital heart disease
C-CHD = Critical congenital heart disease(s)
DAo = Descending aorta
DILV = Double inlet left ventricle
DORV = Double outlet right ventricle
d-TGA = Dextro-transposition of the great arteries
ECG = Electrocardiogram
HLHS = Hypoplastic left heart syndrome
LA = Left atrium
LPA = Left pulmonary artery
LV = Left ventricle
LVOT = Left ventricular outflow tract
L-TGA = Levo-transposition of the great arteries
MPA = Main pulmonary artery
PA = Pulmonary atresia
PDA = Patent ductus arteriosus
PFO = Patent foramen ovale
PLAX = Parasternal long-axis
POS = Pulse oximetry screening
PSAX = Parasternal short-axis
PV = Pulmonary valve
RPA = Right pulmonary artery
RV = Right ventricle
RVH = Right ventricular hypertrophy
RVOT = Right ventricular outflow tract
SAX = Short-axis
SMA = Superior mesenteric artery
TAPVR = Total anomalous pulmonary venous return
TOF = Tetralogy of Fallot
TOF-PA = Tetralogy of Fallot with pulmonary atresia
TV = Tricuspid valve
VSD = Ventricular septal defect

There was also significant geographic variation in rates of prenatal detection across states with a low of only 11%, further reinforcing the need to expand the ability of all sonographers to be able to adequately screen for C-CHD. Lesions identifiable on a 4-chamber view such as atrioventricular canal defect or hypoplastic left heart syndrome have detection rates close to 67%, while those requiring outflow tract visualization such as transposition of the great arteries have considerably lower rates of prenatal detection, ~25%.[5] Prenatal detection rates remain poor for conditions such as total anomalous pulmonary venous return and aortic arch obstruction, due to fetal cardiac physiology and associated challenges with detection.[5-7]

Neonates with C-CHD may present with a variety of findings that would warrant an echocardiogram, including tachypnea, cyanosis, and heart murmurs. However, these may not manifest until after 48 hours of life and therefore may be missed during the newborn hospitalization. This delayed manifestation of symptoms is due to the profound hemodynamic changes that occur in the first few days of life as the neonate transitions from fetal circulation to postnatal circulation. In particular, closure of the ductus arteriosus plays a major role in the hemodynamic deterioration in C-CHD that are ductal dependent for systemic or pulmonary blood flow, and the ductus arteriosus may remain open for days. Delayed or missed diagnosis may result in severe cyanosis and/or cardiovascular collapse after discharge from the hospital, which in turn can result in mortality as well as morbidity from hypoxic-ischemic end organ injury, including neurodevelopmental abnormalities due to brain injury.[8-14] Wren *et al* reported from the United Kingdom that 25% of C-CHD were diagnosed after discharge from the newborn nursery.[14] A United States (U.S.)-based study estimated that 29.5% of live-born infants with non-syndromic C-CHD in the National Birth Defect Prevention Study received a diagnosis more than 3 days after birth and late detection varied by C-CHD type (range 7.5%-62%) as well as geographic site.[15] The newborn hospitalization thus represents a critical window during which screening for and detection of C-CHD could potentially result in improved outcomes for these critically ill neonates.[16] These statistics also demonstrate that a discharged newborn is not necessarily free of C-CHD and needs to be evaluated thoroughly with the development of symptoms.

The purpose of this document is to provide the adult sonographer, who does not typically screen for C-CHD, with the essential information and tools needed to detect C-CHD in newborns and aid in life-saving diagnosis.

Pulse Oximetry for Detection of C-CHD

A common feature of many forms of C-CHD is hypoxemia due to the mixing of oxygenated and deoxygenated blood. Hypoxia has to be quite significant (≥ 4-5 gm/dL of deoxyhemoglobin or an oxygen saturation of ≤ ~80%) for cyanosis to be visible to the naked eye and is particularly difficult to detect in infants with pigmented skin, such as Black or Hispanic infants. Pulse oximetry uses the difference in absorption spectra of wavelengths of light between oxygenated and deoxygenated hemoglobin to detect hypoxemia at much milder levels than those detectable by examination alone and is widely accepted as a noninvasive method to measure oxygen saturation in the blood. Multiple studies have looked into the utility of pulse oximetry screening (POS) to detect C-CHD and normal values in newborns have been reported.[17-23] The American Heart Association and American Academy of Pediatrics issued a joint statement in 2009 presenting the evidence for routine use of pulse oximetry in newborns to detect C-CHD. In an analysis of pooled studies of oximetry assessment performed after 24 hours of life, the estimated sensitivity for detecting C-CHD was 69.6% while specificity was 99%, and the positive predictive value was 47%.[24] False-positive screens that required further evaluation occurred in only 0.05% of infants screened after 24 hours. Subsequently, in 2011, a working group convened with members selected by the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children, the American Academy of Pediatrics, the American College of Cardiology Foundation, and the American Heart Association recommended routine use of POS in well-born and intermediate care nurseries.[25] In September 2011, the U.S. Secretary of Health and Human Services added newborn screening for C-CHD to the Recommended Uniform Screening Panel, an action that was endorsed by academic societies.[26] C-CHD screening with pulse oximetry has become nearly universal in the U.S. with 46 states and the District of Columbia having adopted it into their newborn screening program. A simple algorithm used for POS has been developed to assist the provider in management decisions.[16,27-31]

Targets for Screening

Per the Centers for Disease Control and Prevention (CDC), there are a number of types of C-CHD that are targeted for their reliability of identification by POS. (https://www.cdc.gov/ncbddd/heartdefects/hcp.html#Kemper). They collectively represent common forms of C-CHD presenting with hypoxemia.[30] (Table 1). POS will also detect cyanosis due to a non-C-CHD etiology such as noncritical CHD, sepsis, other infection, persistent pulmonary hypertension, parenchymal or anatomic pulmonary disease, transient tachypnea of the newborn, hypothermia, and hemoglobinopathies.[31] Although not C-CHD, these conditions can pose a significant health risk to the neonate and may

Table 1 POS CDC Targets for C-CHD
d-Transposition of the great arteries
Tetralogy of Fallot
Tricuspid atresia
Truncus arteriosus
Total anomalous pulmonary venous return
Hypoplastic left heart syndrome
Pulmonary atresia
Coarctation of the aorta
Double outlet right ventricle
Ebstein anomaly
Interrupted aortic arch
Single ventricle

need immediate intervention and stabilization. POS may be less effective at identifying obstructive left heart lesions such as aortic valve stenosis and coarctation of the aorta, which are among the congenital lesions at greatest risk for acute cardiovascular compromise; nevertheless, it remains a simple and cost-effective tool to screen for C-CHD.[16]

Impact of a Failed Pulse Oximetry Screening Test

Unlike other newborn screening examinations, a failed POS test mandates immediate evaluation for C-CHD. While physical examination, chest X-ray, and electrocardiography (ECG) can be used to assist with the diagnosis, echocardiography is the diagnostic modality of choice for definitive diagnosis of CHD.[32,33] Specialized equipment (pediatric ultrasound transducers) and machine settings are needed for optimal performance of a neonatal echocardiogram along with interpretation by trained pediatric cardiologists. However, access to pediatric echocardiography and cardiology services may be limited in rural areas and smaller community hospitals. Sometimes, a failed POS screen may result in transfer to a facility where such services are available, thus incurring significant resource utilization while adding anxiety and stress to the family. The need for an echocardiogram of a newborn to be performed and interpreted before discharge has resulted in these studies often being performed by sonographers with limited knowledge and training in pediatric echocardiography and interpretation by adult cardiologists in smaller rural hospitals. Studies have shown that the accuracy of echocardiogram interpretation in pediatric patients by an adult cardiologist is significantly lower than that performed by a pediatric cardiologist.[34,35] In this document, we describe the best practices recommended for use by community sonographers predominantly trained in and practicing adult echocardiography but performing echocardiograms on newborns that have failed POS.

Key Points

- A common feature of C-CHD is hypoxemia leading to cyanosis; however, this can be difficult to detect in infants with pigmented skin.
- Based on recommendations from the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children, as well as the American Academy of Pediatrics, the American College of Cardiology Foundation, and the American Heart Association, there has been an increased push for routine screening of newborns by pulse oximetry screening in the last decade.
- A failed POS mandates immediate evaluation for C-CHD, including echocardiography.
- The purpose of this document is to provide the adult sonographer, who does not typically screen for C-CHD, with the essential information and tools needed to detect C-CHD in newborns and aid in life-saving diagnosis.

RECOMMENDED INFRASTRUCTURE

In order to use echocardiography correctly to screen for congenital heart disease in the newborn, appropriate infrastructure is needed, both at the hospital performing the echocardiogram and at the location of the interpreting pediatric cardiologist. This infrastructure is the same as that needed for an adult echocardiography lab and consists of three major components: age-appropriate echocardiography equipment, a mechanism for storage and transmission of images, and a structured communication process among referring provider, sonographer, and reading physician. However, when performing newborn echocardiograms, there are some additional considerations that will be described below.

Instrumentation and Patient Setting

Echocardiographic equipment used for diagnostic studies should include, at a minimum, hardware and software to perform M-mode and 2D imaging, color Doppler, and pulsed- and continuous-wave Doppler. Newborn echocardiograms are best performed with a variety of probes with a range of frequencies. Mid- to high-frequency transducers (6-12 MHz) should be available for imaging. Near-field imaging in the neonate from the suprasternal, parasternal, and apical views require a high-frequency transducer, typically between 10 and 12 MHz. Anatomy best seen at greater depth (typically from subcostal, apical, and sometimes parasternal windows), as well as color Doppler imaging may require lower-frequency transducers capable of imaging at 6-9 MHz. Additionally, appropriate machine presets should be used for pediatric transducers.

The American Society of Echocardiography Guidelines and Standards for Performance of a Pediatric Echocardiogram recommend the following: 'The video screen and display should be of suitable size and quality for observation and interpretation of all the above modalities. This display should identify the performing institution, appropriate patient identifiers, and the date and time of the study. Range or depth markers should be available on all displays. Measurement capabilities must be present to allow measurement of the distance between two points, an area on the 2D image, blood flow velocities, time intervals, and peak and mean gradients from spectral Doppler studies. Frame rate should be optimized to ensure adequate visualization of anatomy at higher neonatal heart rates.' [36]

The use of electrocardiogram (ECG) leads is a standard part of a neonatal echocardiogram and should be part of every study performed on a newborn when screening for congenital heart disease. The higher heart rate of the newborn makes the ECG tracing particularly important for being able to distinguish phases of the cardiac cycle when carefully reviewing anatomy and blood flow patterns. Ideally, the patient should be placed in a supine position in a darkened room. For suprasternal imaging, gentle extension of the neck is achieved by placing a roll under the shoulders and turning the infant's head slightly to the left. Care must be taken to limit environmental exposure so as to avoid hypothermia and resulting discomfort. This is readily achieved by swaddling the infant and exposing only the windows that are being used for image acquisition. If clinically appropriate, a nurse or physician should be bedside to monitor the patient's oxygen saturation and heart rate. The sonographic gel should be warmed prior to use to help the patient maintain body temperature.

Storage and Transmission of Images

Both the referring hospital (where the echocardiogram is performed) and receiving hospital (where the echocardiogram is interpreted), working in a partnership to screen for congenital heart disease, need to have adequate infrastructure to store images locally and transmit studies between sites. This will typically require involvement of information technology specialists to help set up a process for transmission across the internet. There should be sufficient bandwidth in the connection pathway to transmit studies in a quick and reliable manner, regardless of the time of day. The set-up should allow for images to stream with sufficient speed so as to allow for video clips to play in real time. The process should be streamlined and simple enough for all sonographers to be taught how to transmit studies without assistance, and for all interpreting physicians to be able to reliably access studies. Echocardiography reporting must be standardized in the receiving (interpreting) facility. Provisions must exist for the generation and retention of examination data for all echocardiograms performed. Previous echocardiographic data, images and interpretations must be retrievable for comparison.

All studies should be stored electronically at one or both facilities, although the primary responsibility for storage and archiving rests with the performing facility.

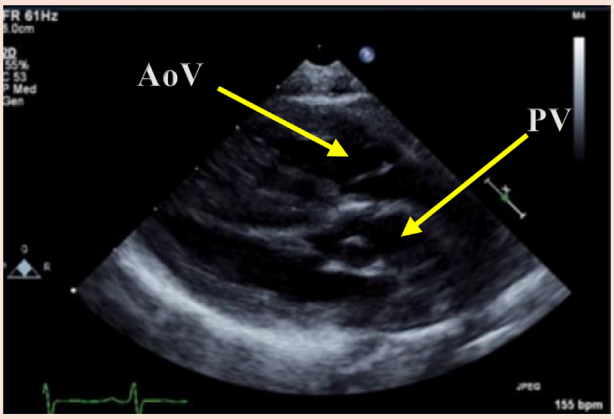
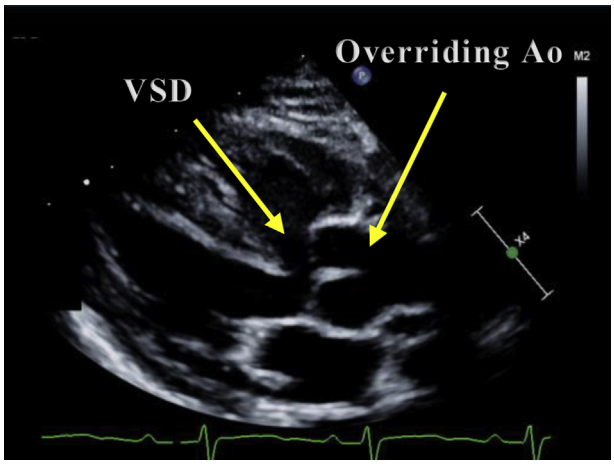
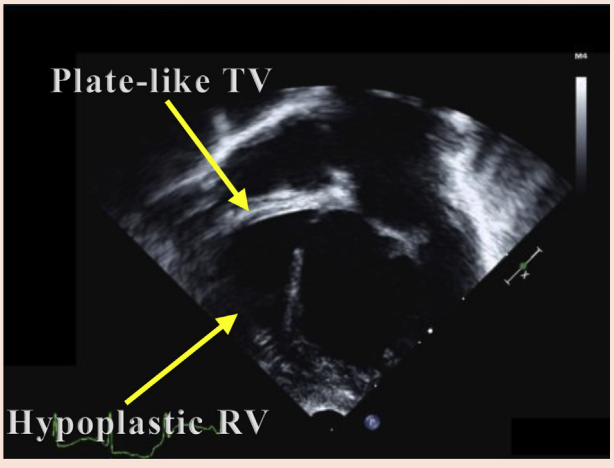
Structured Communication

Hospitals setting up a partnership for screening for congenital heart disease by echocardiography should develop a smooth process for communication. This process begins at the performing site where the newborn nursery or neonatal intensive care unit can notify the receiving site of a pending echocardiogram to review as soon as the

Table 2 Standard & Non-Standard Views for the Adult Sonographer

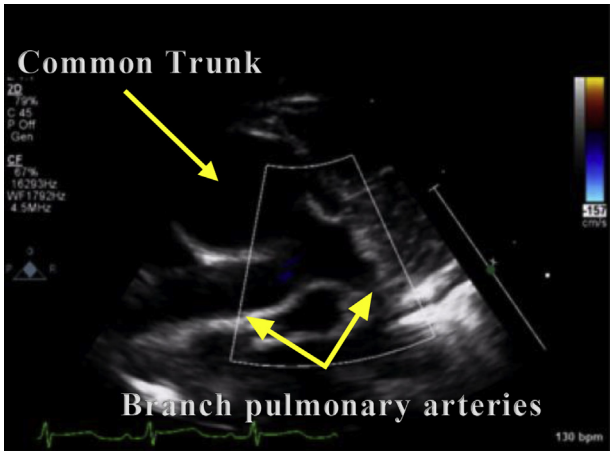
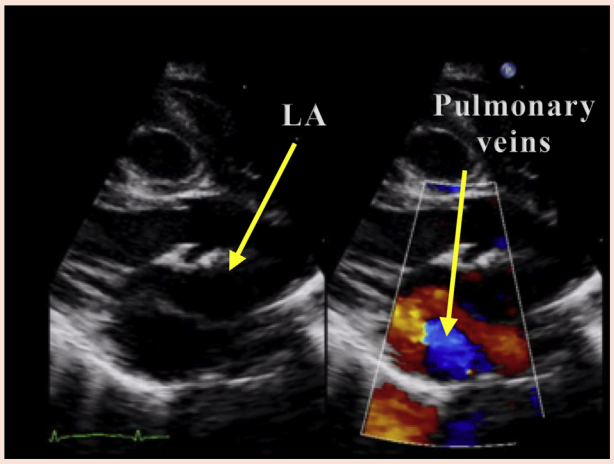
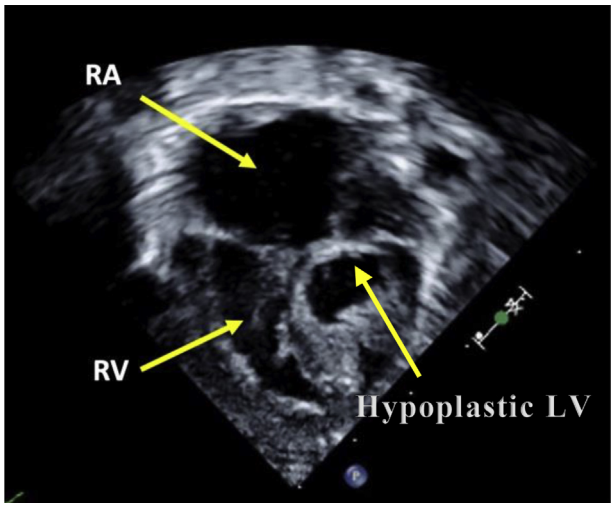
View/sweep	Description	Demonstrated structures	Imaging tips
Standard views			
PLAX sweep	Left sternal border, transducer orientation toward right shoulder, sweeping completely posteriorly and anteriorly	Atrioventricular and semilunar valve orientation, ventricular septum, outflow tracts, ventricular size and function	Sweep slowly through the entire myocardium throughout multiple cardiac cycles.
PSAX sweep	Parasternal window with probe rotated 90 degrees from PLAX view, sweeping from base to apex	Atrioventricular and semilunar valve orientation, pulmonary arteries, ventricular septum, ventricular size and function	
Apical 4-chamber sweep	Probe placed at cardiac apex, sweeping posteriorly to cardiac apex and anteriorly to demonstrate outflow tracts	Atria, ventricles, atrioventricular valves, semilunar valves, outflow tracts, ventricular septum, pulmonary veins	The cardiac apex is not always on the left.
Suprasternal	Long axis	Unobstructed aortic arch	Hyperextend neck (towel roll under shoulder blades, chin up)
Subcostal 4-chamber Sweep (Video 1 available at www.onlinejase.com)	Probe placed in subcostal position, index marker to the right, sweeping posteriorly to anteriorly	2D visualization of all 4 chambers with optimal color and spectral Doppler angle for interrogation of atrial and ventricular level shunting	Image quality may be improved by placing the probe more inferiorly, imaging through the liver.
Non-standard views			
Subcostal SAX (Video 2 available at www.onlinejase.com)	Probe placed in subcostal position, index marker rotated 90 degrees from subcostal 4-chamber view, sweeping from base to apex	2D visualization of all cardiac structures from a SAX cut with optimal angle for color and Doppler interrogation of atrial and ventricular level shunting	
Ductal (Video 3 available at www.onlinejase.com)	High left parasternal sagittal view visualizing the MPA and DAo. If a PDA is present, visualization of the PDA vessel connecting the MPA and DAo	2D visualization of the PDA size and course. Optimal angle for color and spectral Doppler interrogation of PDA shunt direction. Add in sweep from DAo to PA.	Right-to-left ductal shunting can be mistaken for LPA
Abdominal aorta (Video 4 available at www.onlinejase.com)	Subcostal short-axis plane of the abdominal aorta in long axis	Color (demonstrated in Video 4 available at www.onlinejase.com) and spectral Doppler interrogation of the abdominal aortic pulsations. Will demonstrate low-velocity and/or continuous diastolic flow in the setting of proximal obstruction (coarctation).	Angulation of the probe ensuring aortic flow is parallel to the direction of sampling is imperative to obtain accurate spectral Doppler waveforms. Also, important to isolate descending aorta from SMA and celiac artery

Table 3 List of Critical Lesions, Key Findings, and Associated Views

Critical CHD	Echo finding	Echo view
d-TGA  (Video 5 available at www.onlinejase.com)	Side-by-side (parallel) great vessels AoV - anterior & rightward, Pulmonary valve - posterior & leftward PFO L→R shunting MPA arising from LV	PLAX PSAX Subcostal 4-chamber Apical 5-chamber
TOF  (Video 6 available at www.onlinejase.com)	Overriding aorta VSD RVH PDA L→R shunting into branch pulmonary arteries	PLAX Apical 4-chamber High PSAX
Tricuspid atresia  (Video 7 available at www.onlinejase.com)	Plate-like TV Hypoplastic RV RVH PFO R→L shunting	Apical 4-chamber Apical 4-chamber Apical 4-chamber Subcostal 4-chamber

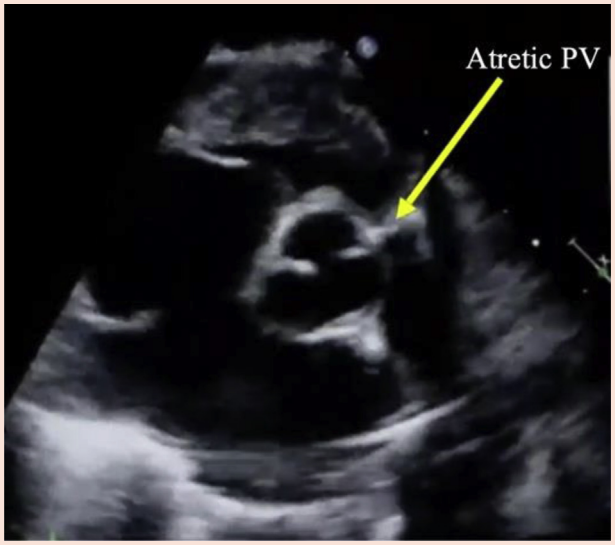
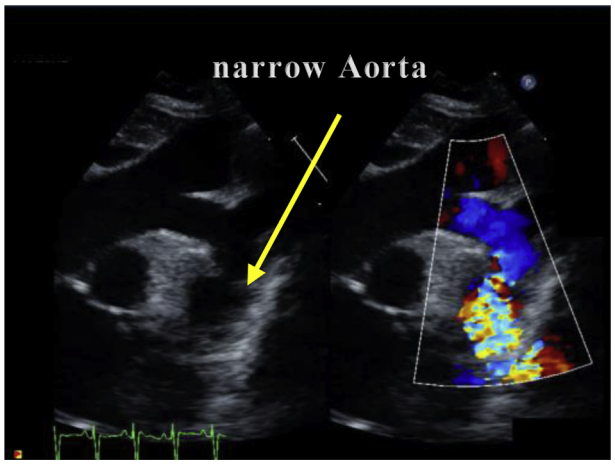
(*Continued*)

Table 3 (*Continued*)

Critical CHD	Echo finding	Echo view
Truncus arteriosus  (Video 8 available at www.onlinejase.com)	Dilated LV VSD/overriding common trunk Pulmonary arteries	PLAX PLAX PSAX, suprasternal
TAPVR  (Video 9 available at www.onlinejase.com)	Dilated RA & RV PFO R→L shunting Small, round LA Posterior pulmonary venous confluence	Apical 4-chamber Subcostal 4-chamber Apical 4-chamber PLAX
HLHS  (Video 10 available at www.onlinejase.com)	Hypoplastic LV Dilated RA & RV PFO L→R shunting	PLAX, PSAX, apical 4-chamber Apical 4-chamber Subcostal 4-chamber

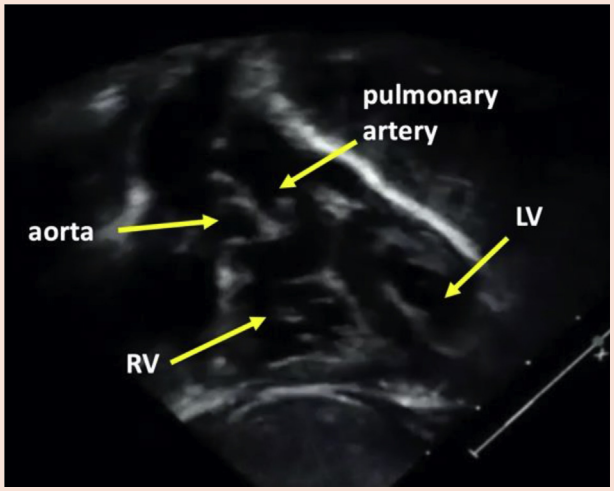
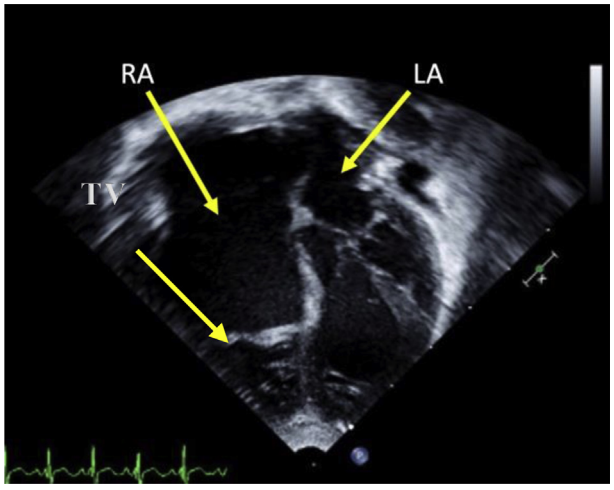
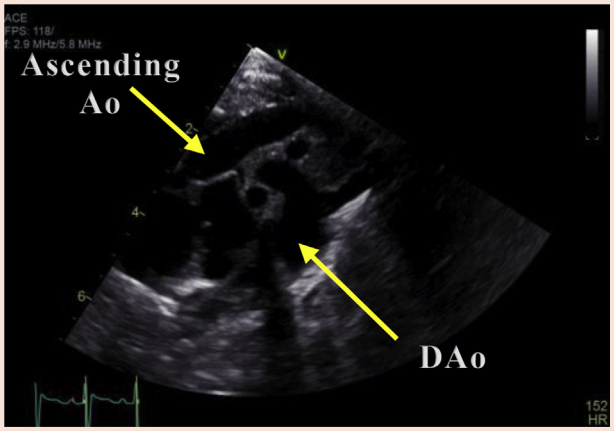
(*Continued*)

Table 3 (*Continued*)

Critical CHD	Echo finding	Echo view
Pulmonary atresia  (Video 11 available at www.onlinejase.com)	No antegrade flow across PV Hypoplastic RV	PLAX, PSAX Apical 4-chamber
Coarctation  (Video 12 available at www.onlinejase.com)	Narrow aorta Diastolic run-off, blunted systolic Doppler pattern	Suprasternal Subcostal short-axis

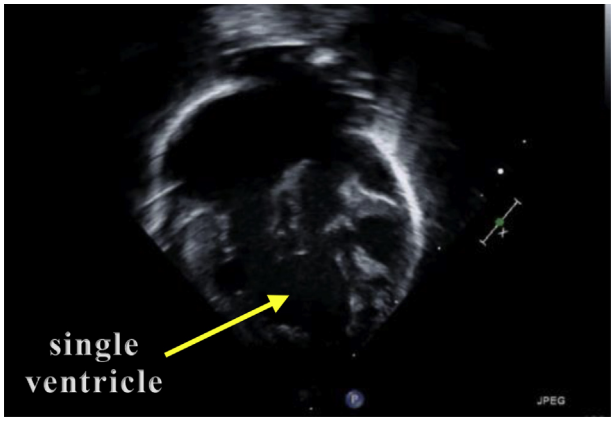
(*Continued*)

Table 3 (*Continued*)

Critical CHD	Echo finding	Echo view
Double outlet right ventricle  (Video 13 available at www.onlinejase.com)	Large subaortic VSD Side-by-side (parallel) great vessels arise from RV Both great vessels arise from the right ventricle	PLAX, apical PLAX, apical Subcostal 4-chamber
Ebstein anomaly  (Video 14 available at www.onlinejase.com)	Apically displaced TV 'Atrialized' RV Possible RVOT obstruction	Apical 4-chamber Apical 4-chamber PSAX
Interrupted aortic arch  (Video 15 available at www.onlinejase.com)	Discontinuity between ascending and descending aorta PDA R→L shunting	Suprasternal PSAX

(*Continued*)

Table 3 (*Continued*)

Critical CHD	Echo finding	Echo view
Single ventricle (DILV)  (Video 16 available at www.onlinejase.com)	Two AV valves connecting to one ventricle	Apical 4-chamber

decision is made to obtain the test. Receiving sites may opt to provide a form (paper or electronic) to performing sites to accompany the echocardiogram being transmitted. Information in this form can include (but is not limited to) demographic information, indication for the study, patient height and weight (for accurate Z-score generation), concurrent systemic blood pressure (for accurate interpretation of pulmonary artery pressure), desired urgency of the interpretation, and contact information so that the study results can be called back to the referring provider. In addition to this information, the referring provider should communicate directly with the reading physician if there is a particular sense of urgency or patient acuity, enabling the reading physician to most effectively interpret the study for the most efficient results and highest quality.

Once studies have been reviewed by a reading physician, results will need to be transmitted back to the performing site securely and efficiently. There must be a policy in place for communicating critical results. This should start with a phone call to the referring provider to relay pertinent results and allow for discussion of patient management if desired. Following this communication, a formal report should be created and finalized, and reports should be returned to the receiving provider by either electronic transmission to the electronic medical record or fax transmission to the inpatient unit. For non-critical results, the hospitals should have an established policy as to whether receipt of the finalized report is considered sufficient communication or if direct provider-to-provider communication is expected on all studies.

Finally, open lines of communication should exist between echocardiography labs at both hospitals. This is important so that sonographers can speak with reading physicians or pediatric cardiac sonographers if they have questions or concerns about a particular study and reading physicians can speak with sonographers to provide feedback and education. Less experienced sonographers are encouraged to speak with the reading physician prior to starting the study to discuss goals and strategies for optimal image acquisition. This two-way communication should be encouraged to continually improve the quality of service given to the referring provider.

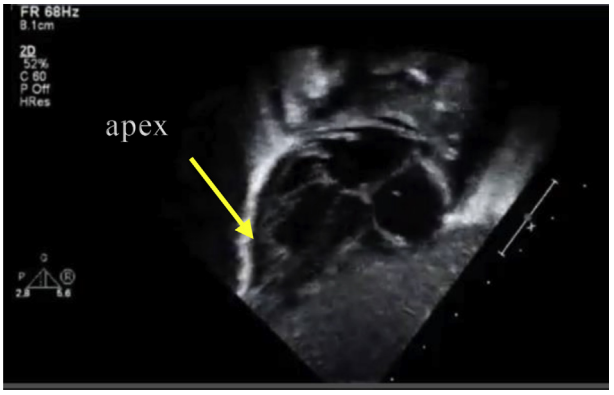
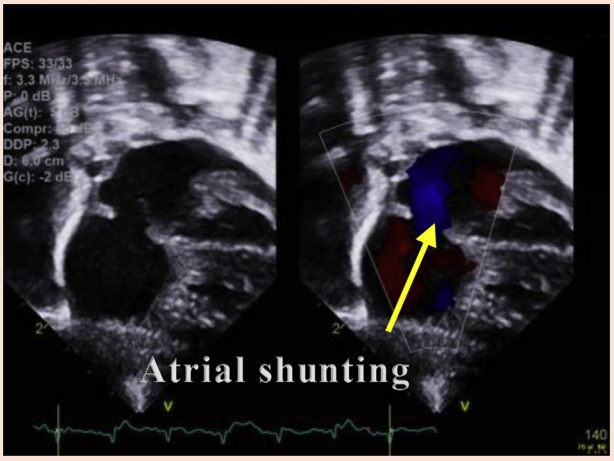
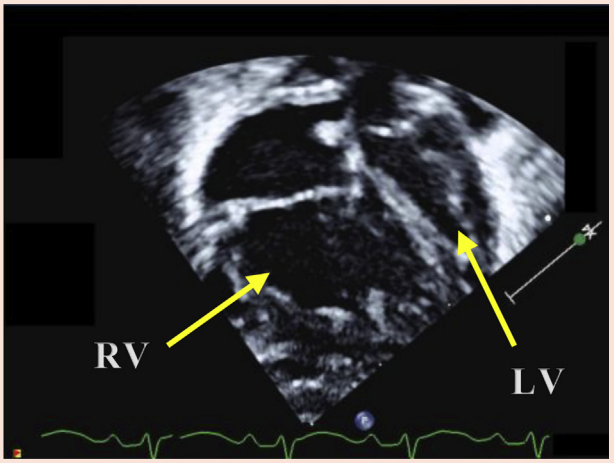
Recommendations

- Centers performing screening echocardiograms in newborns should have a formal relationship with a physician or referral center with expertise in C-CHD.
- These centers should also have available high-frequency transducers, ECG leads, a mechanism for storage and transmission of images, and a structured two-way communication plan.
- The interpreting pediatric cardiologist should work with the referring center to develop a method to relay a final report.

SPECIFIC IMAGING RECOMMENDATIONS

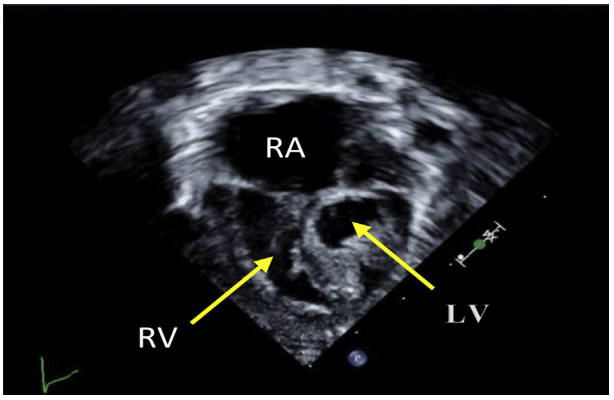
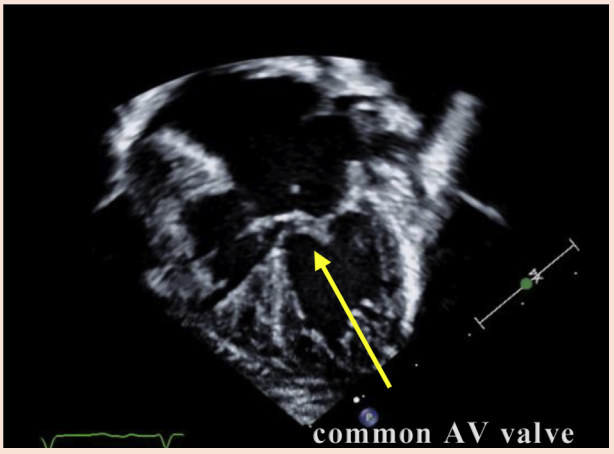
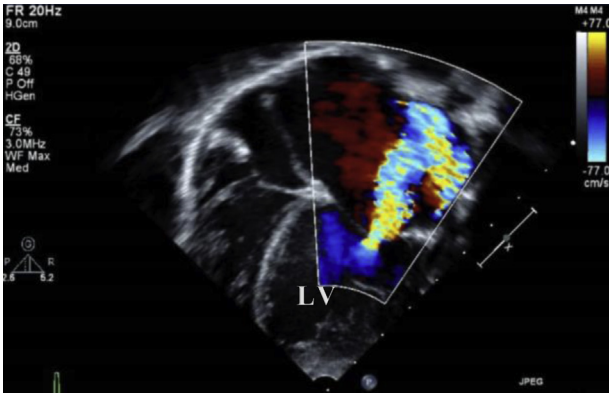
The initial echocardiographic recognition of the presence of C-CHD should be by the imaging sonographer or reading pediatric cardiologist. Therefore, it is recommended that a scanning protocol be developed between the performing and interpreting sites. A standard adult echocardiogram protocol can be followed, as C-CHD can and should be demonstrated in all echocardiographic imaging planes, with the addition of non-standard, traditionally pediatric imaging views and sweeps, deliberately capturing long video clips of data (10-20 seconds). (Table 2). In all imaging views, complete sweeps of the heart should be recorded to rule out abnormalities at its base or apex or at other locations, as well as demonstrate relational orientation of cardiac anatomy. Emphasis on subcostal views is advised as they are generally free from lung artifact and frequently allow for optimal Doppler interrogation of outflow tracts. It is recommended that the sonographer become familiar with pertinent tell-tale echocardiographic findings associated with all forms of C-CHD. (Table 3). Ideally, even if not able to specify the type of C-CHD encountered, the sonographer or echocardiographer should be able to identify 'red flag' findings. (Table 4). Lastly, to facilitate timely diagnosis and appropriate expedited patient care, if C-CHD is suspected on the echocardiogram, the sonographer should stop and notify the local

Table 4 Red Flags in Postnatal Imaging: Differential Diagnosis of Unusual Findings

Echocardiographic findings	Secondary findings	Differential diagnosis
Abnormal Subcostal View		
Abnormal cardiac position  (Video 17 available at www.onlinejase.com)	• Dextrocardia - apex of the heart pointing rightward • Mesocardia - apex is pointing midline	• Complex CHD • Heterotaxy syndromes • Situs inversus totalis
Predominant right-to-left atrial shunt  (Video 18 available at www.onlinejase.com)	• Right-sided obstruction and/or increased right atrial pressure • Little or no blood flowing to the left atrium from the pulmonary veins	• Tricuspid atresia • Pulmonary atresia/intact ventricular septum • Ebstein anomaly • TAPVR
Abnormal Apical 4-Chamber View		
Asymmetry between ventricular sizes  (Video 19 available at www.onlinejase.com)	• Ventricular size discrepancy with otherwise normal structures	• Critical coarctation/aortic arch hypoplasia (larger RV) • TAPVR (larger RV) • Hypoplastic mitral valve

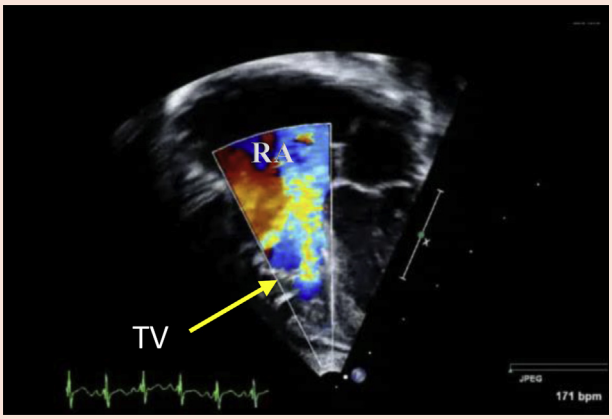
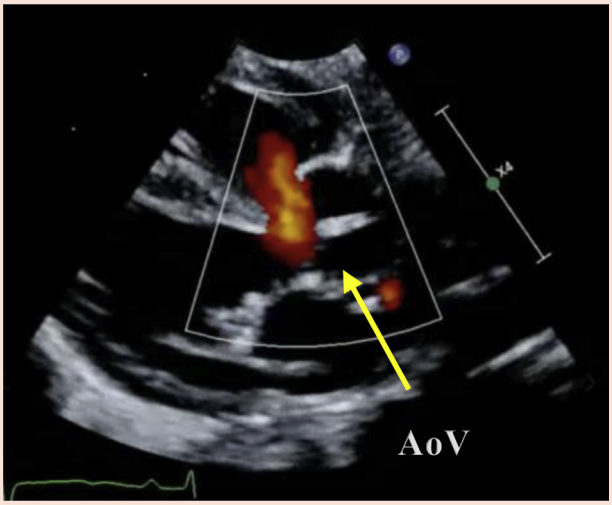
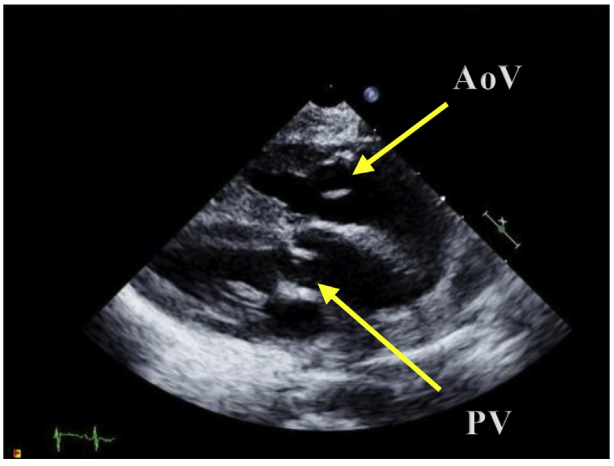
(*Continued*)

Table 4 (*Continued*)

Echocardiographic findings	Secondary findings	Differential diagnosis
Asymmetry between ventricular sizes  (Video 20 available at www.onlinejase.com)	• One ventricle is hypoplastic (non-apex forming) or only one ventricle visualized	• Hypoplastic right heart syndrome • HLHS • Tricuspid atresia • DILV • Single ventricle
Abnormal atrioventricular (AV) valve anatomy  (Video 21 available at www.onlinejase.com)	• Normal appearance of almost equal size mitral and tricuspid valves with slight offset of the tricuspid valve not visualized • One valve is hypoplastic or absent • There is a common AV valve	• Atrioventricular septal defect • Mitral stenosis/atresia • Tricuspid stenosis/atresia • Ebstein anomaly
Marked left AV valve regurgitation  (Video 22 available at www.onlinejase.com)	• Enlarged left atrium • Moderate to severe regurgitation	• Critical aortic stenosis • Cardiomyopathy • l-TGA (because the ventricles are inverted, this would represent "tricuspid" regurgitation)

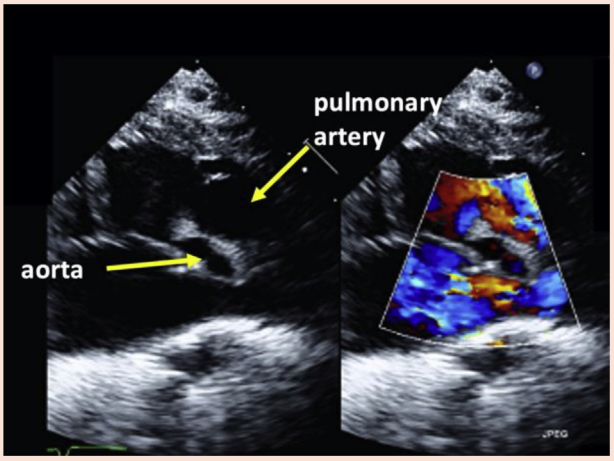
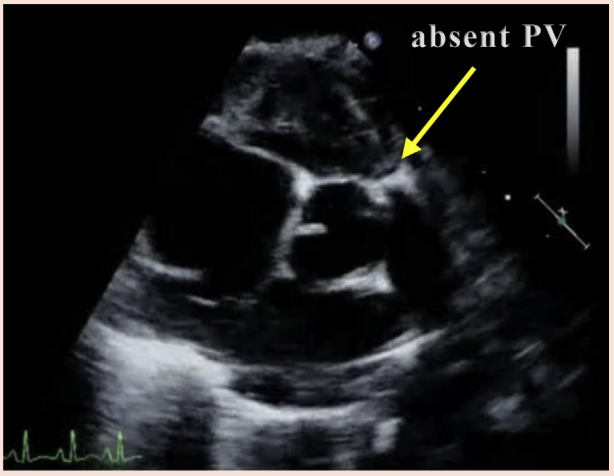
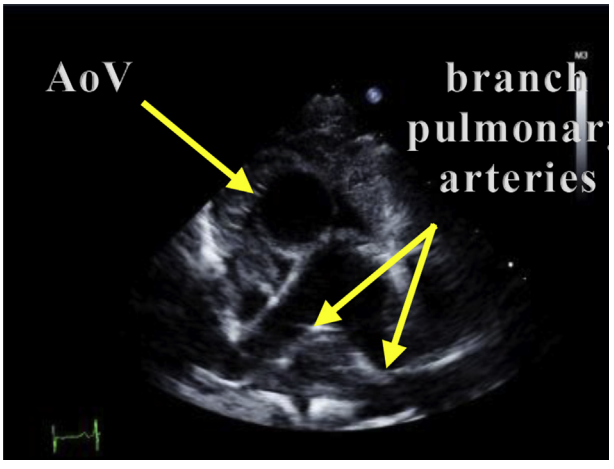
(*Continued*)

Table 4 (*Continued*)

Echocardiographic findings	Secondary findings	Differential diagnosis
Marked right AV valve regurgitation  (Video 23, 24 available at www.onlinejase.com)	• Enlarged right atrium • Moderate to severe regurgitation	• Ebstein anomaly • Cardiomyopathy
Abnormal Parasternal Long-Axis View		
Cannot demonstrate a normal parasternal long-axis view of the left ventricle  (Video 25, 26 available at www.onlinejase.com)	• Left ventricle-to-great vessel relationship is not normal	• TOF • DORV • Truncus arteriosus • Transposition of the great arteries
The orientation of the pulmonary and aortic valves is not normal  (Video 27 available at www.onlinejase.com)	• The pulmonary and aortic outflow tracts are parallel to one another	• DORV • d-TGA • l-TGA (ventricular inversion)

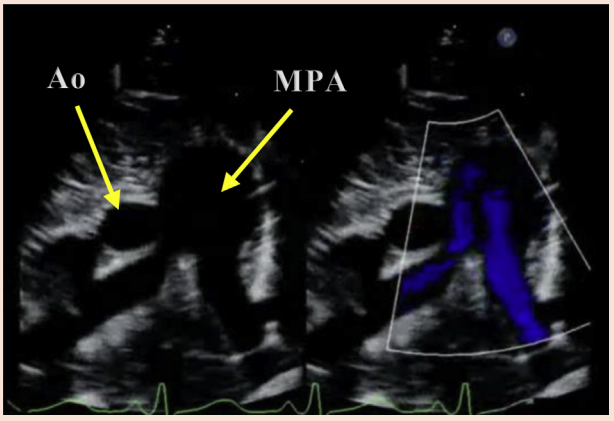
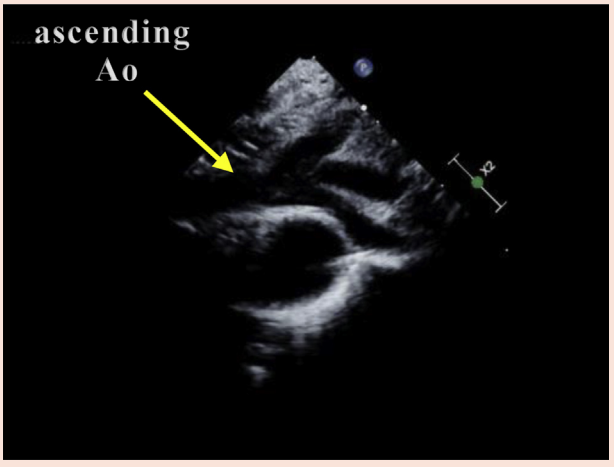
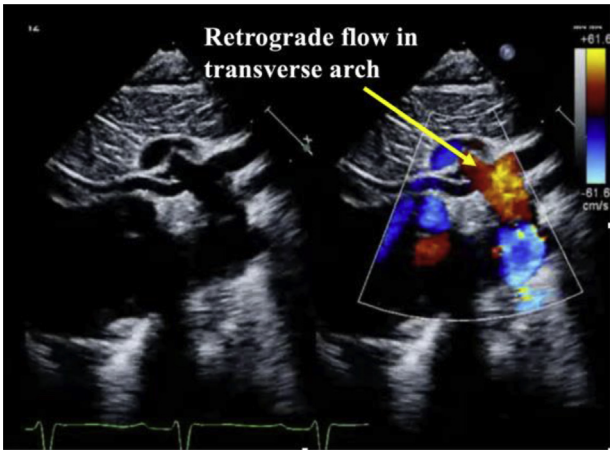
(*Continued*)

Table 4 (*Continued*)

Echocardiographic findings	Secondary findings	Differential diagnosis
Asymmetry between the outflow tracts  (Video 28 available at www.onlinejase.com)	• One outflow tract is significantly smaller than the other or absent	• TOF (small RVOT) • Pulmonary atresia (small RVOT) • Tricuspid atresia (small RVOT) • DORV (variable) • HLHS (small LVOT)
Abnormal Parasternal Short-Axis View		
Cannot demonstrate a normal parasternal short-axis view of the base of the heart demonstrating relationship of RV/PV/MPA  (Video 29 available at www.onlinejase.com)	• Pulmonary valve is small or absent or present and not opening	• Critical pulmonary valve stenosis • TOF with pulmonary atresia/ stenosis • Pulmonary atresia • Truncus arteriosus • Ebstein anomaly (functional pulmonary atresia)
The orientation of the pulmonary and aortic valves is not normal  (Video 30 available at www.onlinejase.com)	• Normal relationship of the aorta and pulmonary artery (pulmonary artery leftward and anterior to aorta) not visualized • The great arteries are side by side, antero-posterior, or the aorta is anterior and rightward/ leftward	• DORV (variable relationship) • d-TGA (aorta right and anterior) l-TGA (ventricular inversion, aorta left and anterior)

(*Continued*)

Table 4 (*Continued*)

Echocardiographic findings	Secondary findings	Differential diagnosis
Size discrepancy between the aortic and pulmonary valves  (Video 31 available at www.onlinejase.com)	• Aortic and pulmonary valves should be almost equal size; one valve is much smaller or absent	• TOF (small PV) • Pulmonary atresia (small PV) • Tricuspid atresia (small PV/AoV) • DORV (variable) • HLHS (small AoV)
Abnormal Suprasternal Notch View		
Inability to lay out aortic arch; cannot obtain the "candy cane" view  (Video 32, 33 available at www.onlinejase.com)	• Small arch structures • Turbulent color flow • Abnormal spectral Doppler pattern	• Coarctation of the aorta • Interrupted aortic arch • Aortic arch hypoplasia • HLHS
Retrograde flow in the ascending aorta and aortic arch  (Video 34 available at www.onlinejase.com)	• Little or no antegrade blood flow across the aortic valve • Retrograde filling of the arch and ascending aorta from the PDA	• Critical aortic stenosis • HLHS • Severe left ventricular dysfunction

(*Continued*)

Table 4 (*Continued*)

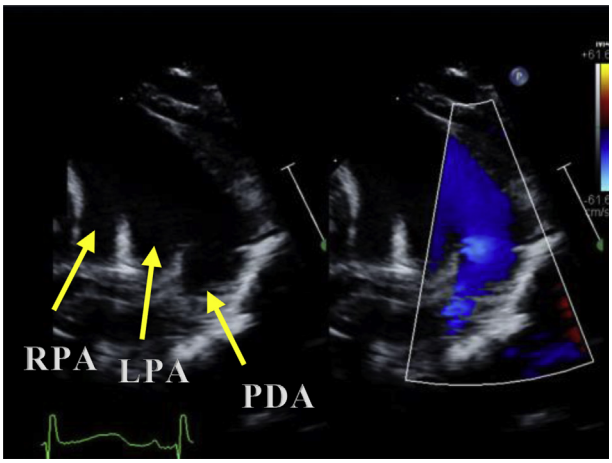
Echocardiographic findings	Secondary findings	Differential diagnosis
Abnormal High Parasternal View (Ductal View)		
Predominant right-to-left flow in ductus arteriosus  (Video 35 available at www.onlinejase.com)	• Blood flow to the descending aorta is coming from the patent ductus arteriosus (PDA)	• Hypoplastic left ventricle • Critical aortic stenosis • Coarctation of aorta • Interrupted aortic arch • Severe pulmonary hypertension
IMPORTANT NOTE: If you can't make it look normal, it probably isn't normal.		

Table showing findings in the neonatal echocardiogram that are suspicious for C-CHD and warrant additional investigation.

provider and/or interpreting pediatric cardiologist before proceeding with the remainder of the study.

Recommendations

- Adult cardiac sonographers performing neonatal screening echocardiograms should commit to learning non-standard, traditionally pediatric imaging views and sweeps to appropriately identify C-CHD.
- The interpreting pediatric cardiologist should work with the referring center to develop a scanning protocol to screen for C-CHD.
- Adult cardiac sonographers should have familiarity with 'red flag' findings and urgently communicate those findings, if present, to their pediatric cardiology partners.

CONCLUSIONS

Timely and accurate diagnosis of C-CHD can improve patient outcomes. With the advent of mandatory POS in newborns prior to discharge from the nursery, community hospitals are increasingly required to perform echocardiograms on newborns to screen for C-CHD. A crucial component to achieving optimal outcomes is the partnership between hospitals performing newborn delivery and care, and offsite pediatric cardiology experts available to interpret newborn screening echocardiography studies, and to assist in the performance of these exams when needed. Specialized equipment, information technology infrastructure, and structured communication are crucial components of the success of these partnerships. By utilizing recommendations contained within this document, it is hoped that more newborns with C-CHD will be accurately identified and stabilized in a timely manner, reducing the incidence of morbidity and mortality in this at-risk population. Additionally, implementation of these recommendations will help sonographers who are not fully trained in pediatric echocardiography to be able to obtain images that allow for accurate diagnosis (or exclusion) of C-CHD.

Reviewers

This document was reviewed by members of the 2020–2021 ASE Guidelines and Standards Committee, ASE Board of Directors, ASE Executive Committee, and designated reviewers. Reviewers included Scott D. Choyce, RDCS, RVT, RDMS, FASE, Meryl Cohen, MD, FASE, David Harrild, MD, FASE, William Katz, MD, FASE, Kelly Thorson, ACS, RDCS, RCCS, FASE, Stephen H. Little, MD, FASE, Anuj Mediratta, MD, FASE, David Orsinelli, MD, FASE, Alan S. Pearlman, MD, FASE, Andrew Pellett, PhD, RDCS, FASE, Geoffrey A. Rose, MD, FASE, Vincent Sorrell, MD, FASE, Madhav Swaminathan, MD, FASE, David H. Wiener, MD, FASE.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.echo.2020.12.005.

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