Consensus recommendations for echocardiography in adults with congenital heart defects from the International Society of Adult Congenital Heart Disease (ISACHD)


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Abstract

The population of adults with congenital heart disease (ACHD) is increasing constantly due to medical, surgical and interventional successes and the input from advanced cardiovascular imaging. ACHD patients are at continuing risk of residua and sequelae related to their CHD contributing to significant morbidity and mortality. Consequently, lifelong expert surveillance is recommended for most patients. Healthcare providers are still working out how best to achieve this objective, how to train enough experts to provide high quality care, and how to organize the delivery of care.

Echocardiography is crucial to clinical surveillance providing a comprehensive assessment of cardiac morphology, physiology, pathophysiology, and function. Thus it contributes significantly to the overall clinical management of ACHD patients. The International Society for Adult Congenital Heart Disease (ISACHD; www.isachd.org) is the leading organization of professionals worldwide dedicated to the pursuit of excellence in the care of ACHD patients. Recognizing the critical role of imaging in the diagnosis and management of ACHD, ISACHD established a task force to provide guidance on echocardiographic studies and reporting. The rationale is that standardization of echocardiographic imaging and reporting carries the potential to improve the overall quality of these exams around the world and facilitate collaborative multicenter research.

The standardized ACHD protocols provided by the ISACHD task force (found in the appendices) include specific recommendations for data acquisition and reporting for each of the major adult congenital heart lesions. These protocols give a comprehensive and structured approach in the evaluation of ACHD patients and help to ensure excellent patient care.

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1. Background

In developed countries, the number of adults with congenital heart defects (ACHD) continues to grow because of historical, surgical, medical, and more recently interventional successes as well as the contribution from other advanced cardiovascular imaging. Indeed, the number of adult patients with CHD now exceeds the number of pediatric patients in many countries [1]. Few have been cured, and most are at continuing risk of residua and sequelae related to their CHD contributing to significant morbidity and mortality. Consequently, lifelong expert surveillance including regular imaging assessment is recommended for most patients [2]. Excellent echocardiography data acquisition and interpretation are essential if excellent patient care is to be provided. Suboptimal echo studies and reports jeopardize patient outcomes. While the needs are often not recognized, these patients also require adult-appropriate diagnostic protocols and expertise for echocardiography applied during surgery, cardiac catheterization and electrophysiology. Diagnostic studies in ACHD also need to differ from both pediatric
cardiac patients and from general adult cardiology patients. More experts in ACHD imaging are needed.

The International Society for Adult Congenital Heart Disease (ISACHD; www.isachd.org) recognizes the critical role of echocardiography in the diagnosis and management of ACHD, and the need to provide guidance on echocardiographic studies and reporting. The rationale is that standardization of echocardiographic imaging and reporting carries the potential to improve the overall quality of these exams around the world and facilitate collaborative multicenter research. Thus, the ACHD echo protocols published in this issue of the International Journal of Cardiology reflect one important step in the maturation of the field of ACHD.

2. Why do we need specific ACHD echocardiography expertise and protocols?

Examples of commonly encountered issues and challenges for echocardiographers based in adult and pediatric echocardiography laboratories are summarized in Table 1. Pediatric echocardiography departments generally benefit from team members who understand CHD and have a high volume of CHD patients. Staff in these laboratories are very familiar with the parameters pertinent to the lesions at hand; the associated lesions and anatomic variants; and the surgical/interventional repairs and their later outcomes. They know what to look for and what they are looking at even when the anatomy is challenging. The staff can, at times, utilize atypical views to better visualize pertinent structures or assess function of a valve or ventricle.

The staff in adult echocardiography departments are typically challenged when imaging CHD patients because of a low volume of CHD patients, especially complex CHD. Usually staff in adult settings are experts in assessing LV function and regional wall motion; the quantitative analysis of valvular function; have regular experience with the use of echocardiography contrast agents and the use of stress echocardiography. In addition, their measurements and reporting are guided by published adult clinical echo guidelines [3–6]. Such guidelines are also available for pediatric patients [7]. The protocols published here reflect the need for this new ACHD echo guidance.

Image orientation from the apical window can be different in adults with CHD compared to those with acquired heart disease and, as a result, adult sonographers may be challenged. Adult sonographers try to apply adult logic and quantitative methods designed for pediatric patients, especially complex CHD. Usually staff in adult settings are experts in assessing LV function and regional wall motion; the quantitative analysis of valvular function; have regular experience with the use of echocardiography contrast agents and the use of stress echocardiography. In addition, their measurements and reporting are guided by published adult clinical echo guidelines [3–6]. Such guidelines are also available for pediatric patients [7]. The protocols published here reflect the need for this new ACHD echo guidance.

Common examples where ACHD echocardiography protocols may benefit both adult and pediatric laboratories.

<table>
<thead>
<tr>
<th>Benefits of ACHD echocardiography protocols</th>
<th>To an adult echocardiography laboratory</th>
<th>To a pediatric echocardiography laboratory</th>
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<tbody>
<tr>
<td>• A low volume of ACHD patients may make the anatomic diagnoses unfamiliar.</td>
<td></td>
<td>• Conventional coronary artery disease assessment is less familiar; e.g. regional wall motion abnormalities defined by the 17 segment model and use of stress echo, both of which require sufficient practice volume for quality assurance</td>
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<tr>
<td>• The sequential segmental analysis approach needed for ACHD patients is not routine.</td>
<td></td>
<td>• Staff may not be familiar with relevant adult guidelines.</td>
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<tr>
<td>• Congenital mitral valve disease may be unfamiliar.</td>
<td></td>
<td>• Degenerative valvular change may be rarely seen. Valve areas are used in adult patients (but not in children), supplementing peak and mean systolic gradients.</td>
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<tr>
<td>• Z scores are not routinely used. While not routinely recommended in these protocols, they may sometimes be useful.</td>
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<td>• Gender-based assessment is less common in pediatric practice though BSA indexing is routine.</td>
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<tr>
<td>• Extrapolation of measurements used in the diseased structurally normal heart may be inappropriate in ACHD patients</td>
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<td>• Contrast echocardiography (seldom used in pediatric laboratories) can be very helpful in evaluating chamber sizes, systolic function, and excluding thrombi in patients with limited acoustic windows.</td>
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<td>• Specific, surgically created structures and their variations such as tunnels, baffles and conduits require specific, unconventional views and familiarity with them.</td>
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<td>• Diastolic dysfunction requires routine assessment in ACHD patients, but not necessarily in children. Right atrial pressure may less commonly be assessed in pediatric practice (cooperation) but is routine for ACHD</td>
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<tr>
<td>• Ventricular function assessment requires appreciation of not only left and right morphology but also their subpulmonary and systemic ventricular position.</td>
<td></td>
<td>• ACHD patients may have received newer treatments e.g. MitraClip, TAVI, TMVR devices and assessment for suitability or complications of these may be less familiar.</td>
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3. General considerations

This document introduces a Digital Acquisition Protocol for Adult Congenital Transthoracic Echocardiography and includes in supplements 12 additional supplementary protocols for 12 individual lesions intended to guide echocardiography studies and data to be included in reports in different types of ACHD patients. The supplementary sections give a comprehensive and structured approach to the evaluation of patients with ACHD that is based on pathology and surgical repair, and include key imaging questions, clinical considerations and technical advice. This will help to ensure important anatomical and functional information is not missed. One of the most important initial steps in imaging a CHD patient is the sequential stepwise approach. Fortunately, once the baseline anatomy and physiology has been described and confirmed this helps dictate the frequency and detail needed at subsequent follow-up. It may not be necessary at follow-up echocardiograms to repeat the full sequential anatomical evaluation in any detail unless there have been intervening therapeutic changes. Follow-up echocardiograms will focus more on functional information with quantitative and/or semi-quantitative assessments.

Echocardiography with all its modalities, including advanced techniques such as real time 3D and speckle tracking, provides comprehensive assessment of cardiac morphology, physiology, pathophysiology and function and contributes significantly to the clinical management of adult patients with CHD. It can provide detailed information about cardiac remodeling and ventricular function following surgical repair or catheter intervention and contributes significantly to clinical management during long-term follow-up. Increasingly, studies have demonstrated the
prognostic value of echocardiography measures in specific ACHD conditions; accordingly, echocardiography can be used for guiding optimal treatment. As the management of CHD continues to evolve, so echocardiography will continue to expand its current applications and remain a pivotal tool in managing adult congenital heart disease patients.

4. Standard digital acquisition protocol for ACHD transthoracic echocardiography (Table 2, Figs. 1–2)

4.1. Image optimization

Standard image optimization techniques should be employed with transducer frequency, image depth, gain and focal zone settings adjusted to optimize image quality. Color flow images should be captured with the highest possible frame rate, aiming for a minimum of 20 Hz using the minimum width necessary. Spectral Doppler images should be appropriately optimized with sample volume size adjusted for flow, choosing the correct baseline, scale and gain settings.

This protocol represents a minimum data set for a standard ACHD echocardiogram. Additions specific to each ACHD lesion such as the use of a non-imaging probe are discussed under lesion-specific protocols appended.

4.2. Measurements

It is recommended that all measurements be performed at the time of recording and stored digitally to allow easy viewing for subsequent reviewers (such as the referring medical team) [8]. It is recommended to also digitally store report pages.

Pediatric echocardiography services favor the use of Z scores which are more infrequently used by ACHD clinicians. The authors are persuaded that Z scores are not routinely required in ACHD echocardiography reports.

4.3. Other considerations

It should also be noted that inclusion of all potential deviations from the standard protocol according to the pathology encountered is outside the scope of this document. Use of off-axis imaging, non-conventional imaging windows and non-imaging transducers is frequently required. In complex pathologies, some standard measurements may not be relevant and may be misleading e.g. septal DTI in the presence of a VSD patch. In other cases, e.g. a systemic right ventricle, standard measurements may have value in serial assessment to follow progression of disease; however, clinically useful lesion specific reference ranges are still not available.

5. Special considerations when assessing ACHD patients with echocardiography

5.1. Ventricular function

5.1.1. Morphologic (sub-aortic) left ventricular function

Conventional methods and standard criteria have been applied for assessing left ventricular global and regional systolic function in the normally connected heart.

Measurement of LV systolic function by shortening fraction assumes a circular geometry with homogenous contraction, conditions that are seldom met in patients with ACHD. The Simpson’s biplane method is preferred when technically feasible. Measurements of LV volumes and EF by 3D echocardiography are preferred over 2D measurements [9]. In patients with a compressed LV due to severe RV dilatation (such as repaired Fallot with severe pulmonary regurgitation), non-geometric indices of longitudinal LV function measured by Tissue Doppler or strain and strain rate measured from speckle tracking have been shown to be associated with outcomes [10].

Although methods and criteria for ventricular diastolic dysfunction are well established for the general cardiac population [11], there is still a lack of reliable parameters for assessing LV diastolic function in ACHD. Criteria used in the general cardiac population such as E/E’ ratio may not be accurate in all congenital heart patients as reduced E’ can be the result of localized surgical scarring in the septum or LV free wall rather than impaired global LV diastolic function [12].
5.1.2. Morphological (sub-pulmonary) right ventricular dimension and function

Echocardiographic assessment of right ventricular (RV) size and function is more difficult, in part because of the shape of the RV [13]. Indirect measures of right ventricular function such as fractional area change (FAC) and tricuspid annular plane systolic excursion (TAPSE) from a four-chamber view are widely used [14, 15] although these techniques exclude assessment of the outflow tract, which plays a major role in some conditions. Tricuspid annular plane systolic excursion represents longitudinal contractile function of the RV, is reproducible, and easy to measure though it may be influenced by tricuspid regurgitation, abnormal ventricular geometry, and recent surgical procedures. Moreover, TAPSE has prognostic value, for example, in Eisenmenger syndrome [16]. Tissue Doppler imaging of myocardial velocities and speckle tracking for myocardial deformation have both been applied in ACHD for regional function.
and global ventricular myocardial deformation assessment but their clinical application to the RV in ACHD remains to be elucidated [10, 17]. 3D echo is now available to measure right ventricular volume and function and can be used to study RV regional as well as global function [18, 19]. RV diastolic dysfunction can be assessed from the presence of pre-systolic forward flow on the pulmonary Doppler “a” wave, by specific hepatic venous flow pattern abnormalities (dominant diastolic flow with increased retrograde flow during atrial contraction) and IVC and right atrial dilatation [20, 21].

5.1.3. Systemic (sub-aortic) right ventricular size and function

There are 2 different congenital anatomic conditions in which the morphological right ventricle supports the systemic circulation: congenitally corrected transposition of the great arteries (ccTGA) and TGA after atrial switch (Mustard or Senning) repair. Because of complex geometry and positional variation, the assessment of systemic RV dimensions and function by echocardiography has remained mostly qualitative. However quantitative parameters including tricuspid plane systolic excursion, fractional area change and tissue Doppler velocity should be routinely used [22–24] for serial assessment.

By analogy with the LV, RV ejection fraction (RVEF) is the marker of RV function. However, the shape of the RV does not allow the use of geometric formulae to calculate the RVEF. Therefore two-dimensional assessment of RVEF has been problematic. FAC measured in the four-chamber view has been shown to be the best conventional variable to correlate with EF measured by CMR [25]. Using a more advanced echocardiography technique – speckle tracking imaging, derived two-dimensional strain and strain rate of systemic right ventricle have been shown to be feasible and associated with adverse clinical outcomes [26, 27]. Real time (RT) three-dimensional echocardiography (3DE), can overcome the problem of geometric assumptions and apical foreshortening and allows accurate acquisition of RV volume and calculated RVEF. Study has shown good correlation between RT3DE and CMR for RVEF in the subpulmonary RV but a large underestimation of RV volumes by echocardiography, especially in severely dilated RVs, with great difficulties in encompassing the entire RV in the pyramidal sector size. Presumably the same limitations would be faced for assessing a systemic RV with this technique.

There is lack of the standard echocardiography criteria for diastolic function in a systemic RV at present. In patients with TGA after atrial switch repair, abnormal filling of the ventricles was not only related to impaired diastolic properties of the ventricles, but also to the filling abnormalities through the noncompliant surgical baffles [28, 29], these decrease the capacity of the systemic venous atrium and diminish the ability of atrial contraction to boost ventricular filling.

5.1.4. Single ventricular function

Systemic ventricular geometry and contractility are significantly different in patients with univentricular physiology compared to patients with normal biventricular physiology [30]. Thus, assessment of ventricular function remains important yet challenging.

Although echocardiography is a mainstay for the serial clinical assessment of ventricular function in this population, standard protocols primarily rely on qualitative assessment due to the complex types of ventricular chamber geometry encountered. Assessment of diastolic dysfunction in this setting is also important but challenging [31–33]. Three-dimensional echocardiography has been shown to be highly feasible in pediatric patients [34, 35].

A recent study has shown that atrioventricular valve systolic to diastolic ratio as a non-geometric parameter correlated well with systemic ventricular end-diastolic pressure and predicted patient outcome [36, 37]. Incorporation of such newer quantitative echocardiographic parameter into clinical practice may aid in identifying abnormalities in ventricular function.

5.1.5. Asynchrony

Echocardiography is the imaging method of choice for detecting asynchrony and thus assists decision-making for pacing and other arrhythmia intervention [38].

5.2. Valve function

Echocardiography is very useful for assessing valvular anatomy and function (e.g. severity of regurgitation and obstruction) and the hemodynamic consequences (atrial or ventricular dilatation and dysfunction) [39–41]. In congenital heart disease, obstruction may also occur at sub- or supra-valvular level such as in parachute mitral valve or tunnel like sub-aortic stenosis. Careful examination by 2D and/or 3D imaging is crucial for accurate diagnosis [42]. Pulmonary valve regurgitation (PR) in repaired tetralogy of Fallot patients and after pulmonary valvotomy for congenital pulmonary stenosis is a common residual lesion with proven detrimental hemodynamic consequences [43]. Quantification of pulmonary regurgitation can be achieved by using 2D, color and Doppler measures (PR index, Pressure half time, M-mode PR index) [44–46]; all of which are needed to improve the accuracy of diagnosis [47].

6. Assessment of specific anatomy including following ACHD intervention

6.1. Coarctation of the aorta

The characteristic Doppler profile across a coarctation is usually diagnostic with a high peak systolic flow velocity and holodiastolic tail. In extremely severe cases, peak flow velocity across the coarctation may be very low, but flow becomes continuous. On the other hand, the severity of a long tubular segmental narrowing may not be accurately estimated by the modified Bernoulli equation. Flow in the abdominal aorta shows reduced pulsatility and increased diastolic component in significant coarctation with established collaterals [48].

6.2. RV to PA conduits

Creation of an extracardiac RV to PA conduit is often required in patients with pulmonary atresia or transposition of great arteries with severe pulmonary stenosis. The position of these conduits are frequently anterior, some of them immediately behind the sternum, therefore unconventional views are needed. Color flow mapping is helpful in identifying their location and the continuous wave Doppler probe can be used to detect the peak flow velocity [49]. The pressure gradient calculated by the modified Bernoulli equation using peak flow velocity may not be accurate if there is a long tubular narrowing.

6.3. Venous pathways in transposition of great arteries after atrial redirection surgery (Mustard and Senning operation)

Venous pathway obstruction can be assessed using 2D, color flow mapping and Doppler. Low-scale (lowering the Nyquist limit) color Doppler should be used. Given imaging is of a venous pathway, flow velocity >1.6 m/s is suggestive of pathway obstruction. In severe obstruction, flow can become continuous with very low or even undetectable velocity with Doppler. Venous pathway (baffle) leak is common, and mixed saline contrast is useful in detecting the site and severity of the shunt.

6.4. Branch pulmonary arteries for transposition of the great arteries after arterial switch repair

Branch pulmonary artery stenosis is one of the common complications after arterial switch repair for TGA especially when the LeCompte maneuver has been used [50]. The obstruction is usually very difficult to
visualize on standard 2D views. High parasternal and suprasternal views are often needed. In these patients, increased tricuspid regurgitation flow velocity can be an indirect sign of distal pulmonary stenosis which will more frequently be the explanation than increased pulmonary vascular resistance, although the latter can also occur.

7. Conclusion
The strengths of echocardiography include its ease-of-use, portability, and accuracy in assessing cardiac anatomy and function. Consequently, it is the most commonly used tool for diagnosis and follow-up of ACHD patients. Echocardiography with all its modalities and advanced techniques such as real-time 3D and speckle tracking provides a comprehensive assessment of cardiac morphology, physiology, pathophysiology, and function – and thus contributes significantly to the clinical management of adult patients with CHD. Echocardiography can provide detailed information regarding cardiac remodeling and ventricular function following surgical repair or catheter intervention. Echocardiographic data has prognostic significance. It is thus extremely important that sonographers and echocardiographers alike have a solid knowledge and a specific approach to data acquisition and reporting for the major categories of congenital heart defects seen in adult patients. The protocols provided in this issue of the International Journal of Cardiology provide recommendations to guide the performance of echocardiograms on ACHD patients performed in either pediatric or adult laboratories.

Acknowledgements
Dr. Erwin Oechslin currently holds the Bitove Family Professorship of Adult Congenital Heart Disease.
Dr. Sonya V Babu-Narayan is supported by the British Heart Foundation [FS/11/38/28864].

Conflict of interest
The authors report no relationships that could be construed as a conflict of interest.

Appendix A. Supplementary data
Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijcard.2018.07.058.

References