

Congenital anatomy, acquired pathology – A synergistic approach to echocardiographic evaluation of the adult with congenital heart disease

1 | INTRODUCTION

Echocardiographic evaluation of an adult with congenital heart disease (ACHD) must thoroughly address both underlying anatomic complexity and acquired pathology. For a complete assessment in these complex cases, the ACHD practitioner benefits from techniques developed in both adult and pediatric echocardiography laboratories.

The strengths of the two laboratories can be complementary for the ACHD practitioner. Pediatric echocardiographic examination is tailored for the anatomic characterization of congenital heart diseases, including changes from birth and somatic growth. Invasive structural palliations may be necessary in childhood, and echocardiographic examination includes evaluation of shunts and conduits when necessary. Adult echocardiographic examination, by contrast, is tailored for acquired heart diseases whose prevalence largely increase with age, such as ischemic heart disease and calcific aortic stenosis. While adults may undergo invasive procedures, anatomic factors such as cardiac situs and chamber orientation are generally assumed to fit a consistent pattern.

Consensus recommendations from the International Society of Adult Congenital Heart Disease¹ acknowledge that ACHD echocardiography expertise is unique, and requires drawing from the strength of each laboratory. In practice, though, combining the approaches can be challenging.

2 | STRENGTHS OF ADULT ECHOCARDIOGRAPHY LABORATORIES

The adult echocardiography protocol offers important tools for the quantification and grading of acquired conditions. The valve guidelines in adult echocardiography^{2,3} showcase the strength of quantification techniques. The use of the continuity equation to estimate valve area in aortic valve stenosis is a well-known example⁴; to underline the strength in quantification, no fewer than 11 additional methods of quantifying aortic stenosis severity are included in the guidelines.

The adult echocardiography laboratory also excels in the evaluation of acquired insults. An evaluation of left ventricular function, for

example, accounts for the possibility of coronary artery disease with regional wall motion analysis. Types of left ventricular remodeling are also accounted for, including global and regional changes seen in a variety of acquired conditions.⁵ Adults with congenital heart disease, many also at risk for commonly acquired conditions,⁶ benefit from this focus.

Challenging acoustic windows are commonly encountered in adult laboratories. This obstacle has given rise to useful adaptations, such as routine use of echocardiographic contrast and a lower threshold to refer for transesophageal echocardiography.

3 | STRENGTHS OF PEDIATRIC ECHOCARDIOGRAPHY LABORATORIES

The anatomic complexity of certain congenital conditions demands a different approach, which shows the strength of the pediatric laboratory. The 17-segment model of the left ventricle would not be applicable to a patient with hypoplastic left heart, for example. Techniques for quantifying aortic valve stenosis may also encounter new inconsistencies if the aortic valve arises from a *right* ventricular outflow tract; the angles for optimal Doppler interrogation would be different, and outflow tract diameter calculations would be less consistent in a more oblong structure. In the pediatric age group, the large variation in body and heart size adds an additional challenge. Because of these differences in pathology, adult valve quantification methods cannot simply be applied to a pediatric age group.⁷

The guideline for performance of a pediatric echocardiogram specifies that the approach is designed for the “wide spectrum of anomalies encountered in patients with congenital heart disease.”⁸ Echocardiographic views which would be unconventional in an adult laboratory, as well as probe “sweeps” take on added importance in this setting. Additionally, somatic growth and the large variability of body sizes demand that all essential measurements be expressed relative to body size.⁹

4 | STRENGTH THROUGH TECHNICAL SYNTHESIS IN THE ACHD LAB

Echocardiography in ACHD provides a unique opportunity to harness the strengths of both adult and pediatric echocardiographic techniques. Evaluation of the systemic right ventricle (sRV) provides an

Abbreviations: ACHD, adult congenital heart disease; ccTGA, congenitally corrected transposition of the great arteries; d-TGA, dextro-transposition of the great arteries; sRV, systemic right ventricle.

instructive example. Recognition of conditions in which sRV is present – such as congenitally corrected transposition of the great arteries (ccTGA) or d-transposition of the great arteries (d-TGA) palliated with atrial switch operation – is the first step, and requires anatomic imaging expertise which is the forte of the pediatric laboratory.¹⁰ sRV also represents a degenerative condition, and this makes quantification of ventricular function, a major focus in the adult lab, crucial for effective clinical care. Studies demonstrating the utility of global longitudinal strain^{11,12} and fractional area change^{13,14} for the evaluation of sRV function typify effective synergy of adult and pediatric approaches to assess complex pathology.

5 | IMPLICATIONS FOR THE FUTURE OF ECHOCARDIOGRAPHY IN ACHD

Effective ACHD imaging depends on a successful synergy of anatomic information and functional quantification, bringing together techniques from pediatric and adult echocardiography laboratories.

- *Acknowledgment of anatomic heterogeneity* is necessary to account for the breadth of conditions encountered. Techniques from pediatric echocardiography guidelines should be employed for anatomic evaluation, such as the determination of visceral situs and the use of sweeps.
- *Familiarity with structural (both surgical and transcatheter) palliations* for congenital heart disease is essential. When an intervention is anticipated, necessary data for planning should be obtained. Close collaboration between sonographers and physicians is strongly encouraged.
- Attention must be paid to the possibility of *multiple acquired insults*. In addition to chamber sizes (indexed to BSA when appropriate), evaluation of regional ventricular wall motion should be included in any adult examination, and careful interrogation of each valve is necessary to evaluate for new or progressive disease.
- *Quantification techniques* are essential for gathering reproducible, valid data. When possible, the range of established quantitative methods commonly used in adult laboratories, such as the estimation of valve orifice area in aortic stenosis or mitral regurgitation, should be applied.
- *Poor acoustic windows should be anticipated*. Echocardiographic contrast media or transesophageal echocardiography should be used when necessary to see anatomic structures with an appropriate level of detail.

Future research should expand the validation and use of quantification techniques in adult congenital heart disease. The diagnostic imperative for evaluation of the ACHD patient will be the development of a collaborative “hybrid” echocardiography laboratory which can provide more than just part of the data necessary for care.

CONFLICTS OF INTEREST

Nothing to disclose.

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