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ORIGINAL ARTICLE

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Hand-held echocardiography in children with hypoplastic left heart syndrome

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Abstract

Background: When performed by cardiologists, hand-held echocardiography (HHE) can assess ventricular systolic function and valve disease in adults, but its accuracy and utility in congenital heart disease is unknown. In hypoplastic left heart syndrome (HLHS), the echocardiographic detection of depressed right ventricular (RV) systolic function and higher grade tricuspid regurgitation (TR) can identify patients who are at increased risk of morbidity and mortality and who may benefit from additional imaging or medical therapies.

Methods: Children with HLHS after Stage I or II surgical palliation (Norwood or Glenn procedures) were prospectively enrolled. Subjects underwent HHE by a pediatric cardiologist on the same day as standard echocardiography (SE). Using 4-point scales, bedside HHE assessment of RV systolic function and TR were compared with blinded assessment of offline SE images. Concordance correlation coefficient (CCC) was used to evaluate agreement.

Results: Thirty-two HHEs were performed on 15 subjects (Stage I: n = 17 and Stage II: n = 15). Median subject age was 3.4 months (14 days-4.2 years). Median weight was 5.9 kg (2.6-15.4 kg). Bedside HHE assessment of RV systolic function and TR severity had substantial agreement with SE (CCC = 0.80, CCC = 0.74, respectively; $P < .001$). HHE sensitivity and specificity for any grade of depressed RV systolic function were 100% and 92%, respectively, and were 94% and 88% for moderate or greater TR, respectively. Average HHE scan time was 238 seconds.

Conclusions: HHE offers a rapid, bedside tool for pediatric cardiologists to detect RV systolic dysfunction and hemodynamically significant TR in HLHS.

KEYWORDS

congenital heart disease, focused cardiac ultrasound, hypoplastic left heart syndrome

1 | INTRODUCTION

Hand-held echocardiography (HHE) platforms developed over the past decade have shown promise as an "ultrasonic stethoscope" available to augment a clinician's bedside physical examination.[1-3] However, shallow imaging depths, small rib spaces, and the need for high temporal and spatial resolution in small children have limited the utility of HHE in the pediatric population. The largest published HHE pediatric series are in rheumatic heart disease screening programs in children with structurally normal hearts, and the use of HHE in congenital heart disease in available literature is mostly limited to simple septal defects and two ventricle circulations.[4-8]

Infants with hypoplastic left heart syndrome (HLHS) are at high risk for clinical deterioration and death, particularly in the interstage period between their initial Norwood palliation and Stage II Glenn procedure.[9] The early detection of depressed right ventricular systolic function and higher grade atrioventricular valve regurgitation by echocardiography can assist in identifying patients with HLHS who are at increased risk for morbidity and mortality and would benefit from further study and/or intervention.[9-12] The bedside availability of HHE makes it an attractive tool in the clinical management of this high-risk patient population. In this article, we present a pilot study evaluating the feasibility of pediatric cardiologists using HHE to identify right ventricular systolic dysfunction and/or hemodynamically significant TR in children with HLHS.

2 | METHODS

This prospective, single-center study was designed to assess the accuracy of bedside HHE use by a pediatric cardiologist to identify right ventricular (RV) systolic dysfunction and significant tricuspid regurgitation (TR) in infants and children with HLHS. Focused HHEs did not include other imaging, such as, evaluation of the aortic arch or shunt patency.

Local institutional review board approved the study design prior to enrollment and written informed consent was obtained from all participants' guardians.

HHE imaging was performed with a General Electric Healthcare Vscan portable ultrasound system (GE Vscan). The device measures 13.5 × 7.3 × 2.8 cm and weighs ~390 g with the probe included. The display screen measures 5.3 × 7.1 cm with a 3:4 aspect ratio and a resolution of 240 × 320 pixels. Grey scale and color Doppler modalities are available. There is a single-phased array probe with a frequency range between 1.7 and 3.8 MHz and a footprint measuring 3.3 × 2.6 cm. Still frames, cine loops, and voice recordings can be stored on a 4 GB micro-SD memory card in MPEG-4 format and can later be uploaded to a computer hard drive for off-line interpretation. Each cardiologist underwent a brief introductory training session (run by A.R.), which included familiarization with device controls and strategies for image optimization. Cardiologists each trialed the device prior to the start of the study until they reported feeling comfortable with the controls.

Patients with HLHS who had undergone Stage I (Norwood) or II (Glenn) surgical palliations were enrolled in this study. Participants underwent focused, bedside HHEs by a board certified pediatric cardiologist on the same day that standard echocardiograms (SE) were obtained for clinical indications. Participants were recruited and HHEs were performed on a convenience basis depending on investigator and subject availability. The cardiologist performing the bedside HHE was blinded to the results of the SE. HHEs were performed by three pediatric cardiologists (A.R., E.O., R.L.); two (A.R., R.L.) were staff cardiologists in the echocardiography laboratory and one (E.O.) was a cardiologist who attended in the cardiovascular intensive care unit.

SE images were stored in standard DICOM (Digital Imaging and Communications in Medicine) format for later review. HHE interpretation was performed at the time of image acquisition and was based on qualitative 4-point scales for grades of TR and RV systolic function. De-identified HHE images were then uploaded and stored in a secured computer hard drive in MPEG-4 format. SE studies were analyzed offline at a later time by an echocardiographer (A.R., R.L.), who did not perform the HHE and was blinded to results of the HHE. TR and RV systolic function on SE images were analyzed on the same 4-point scales. During SE and HHE assessments, grade of TR was based on qualitative Doppler imaging (color flow jet area and flow convergence zone).[13] For the 4-point scale, assessment of RVSF during SE and HHE was qualitative and based on expert "eyeball method." RV fractional area change was also manually measured on the SE images by the same blinded pediatric echocardiographer.[14] Sensitivity, specificity, and the concordance correlation coefficient (CCC) for repeated measures for interobserver reliability were calculated.

3 | RESULTS

Fifteen subjects (5 females, 10 males) were enrolled and there were 32 encounters (HHE and SE performed on the same day) over a one-year period. The median subject age at time of encounter was 3.4 months (Table 1). Seventeen encounters occurred after Stage I surgical palliation and 15 encounters occurred after the Stage II surgical palliation; eight subjects had repeat encounters during the study period.

The HHEs were performed in the inpatient ward (n = 7), cardiovascular intensive care unit (n = 12), and outpatient clinic (n = 13). The average HHE scan time was 238 seconds.

SE identified depressed RV systolic function in 5 subjects (6 encounters: mild—1, moderate—3 and severe—2) and moderate or greater TR in seven subjects (16 encounters: moderate—13 and severe—3) (Table 2). RV fractional area change ranged from 10% to 56% (median: 36%). All six SE's with depressed RV systolic function

TABLE 1 Subject characteristics (n = 15)

Variable (at time of encounter)	Range
Age	14 days to 4 years old
Gender	Male: n = 10
	Female: n = 5
Weight	2.6-15.4 kg
Height	49-102 cm
Anatomy	MA/AA: n = 8
	MS/AS: n = 5
	MS/AA: n = 2
Stage I surgical palliation	BTTS: n = 7
	Sano: n = 8

Abbreviations: AA, Aortic atresia; AS, Aortic stenosis; BTTS, Blalock-Thomas-Taussig Shunt; MA, Mitral atresia; MS, Mitral stenosis.
Sano: Right ventricle to pulmonary artery conduit.

TABLE 2 Comparison of subject's right ventricular systolic function and tricuspid regurgitation by bedside HHE echocardiogram versus standard echocardiography imaging

Subject		Encounter #1		Encounter #2		Encounter #3		Encounter #4		Encounter #5	
#1		*s/p Norwood POD #14*		*s/p Norwood POD #29*							
		HHE	*S. Echo*	*HHE*	*S. Echo*						
	RVSF	Normal	Normal	Normal	Normal						
	TR Grade	Moderate	Severe	Moderate	Severe						
#2		*s/p Norwood POD #164*		*s/p Glenn POD #77*		*s/p Glenn POD #133*					
		HHE	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*				
	RVSF	Normal	Normal	Mild	Moderate	Mild	Normal				
	TR Grade	Moderate	Moderate	Moderate	Moderate	Moderate	Moderate				
#3		*s/p Glenn POD #42*		*s/p Glenn POD #129*							
		HHE	*S. Echo*	*HHE*	*S. Echo*						
	RVSF	Mild	Normal	Moderate	Severe						
	TR Grade	Severe	Moderate	Moderate	Moderate						
#4		*s/p Norwood POD #18*		*s/p Norwood POD #83*		*s/p Norwood POD #97*		*s/p Glenn POD #7*			
		HHE	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*		
	RVSF	Normal	Normal	Normal	Normal	Normal	Normal	Mild	Moderate		
	TR Grade	Mild	Mild	Trivial	Trivial	Mild	Mild	Trivial	Mild		
#5		*s/p Norwood POD #18*		*s/p Norwood POD #34*		*s/p Norwood POD #48*		*s/p Norwood POD #55*		*s/p Glenn POD #128*	
		HHE	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*
	RVSF	Normal	Normal	Normal	Normal	Normal	Normal	Normal	Normal	Normal	Normal
	TR Grade	Moderate	Moderate	Severe	Moderate	Moderate	Moderate	Moderate	Moderate	Mild	Mild
#6		*s/p Norwood POD #84*		*s/p Glenn POD #148*							
		HHE	*S. Echo*	*HHE*	*S. Echo*						
	RVSF	Normal	Normal	Moderate	Normal						
	TR Grade	Trivial	Trivial	Mild	Mild						
#7		*s/p Glenn POD #1540*									
		HHE	*S. Echo*								
	RVSF	Normal	Normal								
	TR Grade	Trivial	Trivial								
#8		*s/p Norwood POD #16*									
		HHE	*S. Echo*								
	RVSF	Normal	Normal								
	TR Grade	Moderate	Moderate								

(Continues)

TABLE 2 (Continued)

Subject		Encounter #1		Encounter #2		Encounter #3		Encounter #4		Encounter #5
#9		*s/p Glenn POD #826*								
		HHE	*S. Echo*							
	RVSF	Normal	Normal							
	TR Grade	Mild	Mild							
#10		*s/p Norwood POD #4*		*s/p Norwood POD #76*		*s/p Glenn POD #0*				
		HHE	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*			
	RVSF	Normal	Normal	Moderate	Moderate	Severe	Severe			
	TR Grade	Severe	Moderate	Moderate	Severe	Mild	Moderate			
#11		*s/p Glenn POD #338*								
		HHE	*S. Echo*							
	RVSF	Normal	Normal							
	TR Grade	Moderate	Mild							
#12		*s/p Norwood POD #41*		*s/p Norwood POD #55*		*s/p Norwood POD #83*		*s/p Glenn POD #160*		
		HHE	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*	*HHE*	*S. Echo*	
	RVSF	Normal	Normal	Normal	Normal	Normal	Normal	Mild	Mild	
	TR Grade	Mild	Mild	Mild	Mild	Moderate	Mild	Moderate	Moderate	
#13		*s/p Glenn POD #43*								
		HHE	*S. Echo*							
	RVSF	Normal	Normal							
	TR Grade	Trivial	Mild							
#14		*s/p Glenn POD #727*								
		HHE	*S. Echo*							
	RVSF	Normal	Normal							
	TR Grade	Mild	Trivial							
#15		*s/p Glenn POD #325*								
		HHE	*S. Echo*							
	RVSF	Normal	Normal							
	TR Grade	Mild	Mild							

based on the 4-point scale had fractional area change of less than 30%; no patient with fractional area change greater than 30% had depressed RV systolic function based on the 4-point scale.

Bedside HHE assessment of RV systolic function and TR severity had substantial agreement with SE (CCC = 0.80, CCC = 0.74, respectively; $P < .001$). The sensitivity and specificity for detection of any grade of depressed RV systolic function by HHE were 100% and 88%, respectively (Figure 1). The sensitivity and specificity for detection of moderate or greater TR by HHE were 94% and 88%, respectively (Figure 2).

4 | DISCUSSION

We performed a prospective pilot study to evaluate the agreement between HHE and SE in children with HLHS, and we found substantial agreement between the two modalities on the 4-point scales. In the dichotomous evaluation of RVSF and TR, bedside use of HHE by a cardiologist performed excellently and was able to help determine simply, in the vast majority of encounters, whether the RV systolic function was normal or not, and whether or not there was at least moderate TR.

FIGURE 1 HHE Black and white imaging via pediatric apical view; Subject #6 at 5-month old; to obtain this "pediatric friendly" view at the bedside, the entire device is turned upside down and the dot of the probe is inverted toward subject's right shoulder. This image was inverted for publication purposes, but represents what the cardiologist would have seen at the bedside

The clinical utility of HHE in pediatric patients has not been established. However, our small series demonstrates the potential for HHE as an effective screening point-of-care device for a cardiologist during focused evaluations of small children and infants with single ventricle congenital heart disease. Previous HHE studies have shown that its use at bedside can significantly increase the diagnostic capabilities of a cardiologist's evaluation beyond their cardiac physical examination. In an adult cohort, Mehta et al showed that the use of HHE-augmented physical examination increased the accuracy of diagnosing left ventricular dysfunction from 35% to 96% when compared to physical examination alone.[15] In school age and adolescent children, the sensitivity for detection of definitive rheumatic heart disease (based mostly on pathologic mitral or aortic valve disease) increased from ~22% by auscultation alone to ~98% when HHE was utilized.[5] While we did not compare HHE-augmented evaluation to physical examination alone, this is the first study, to our knowledge, to explore the utility of HHE in a cohort of infants and young children with single ventricles.

Over the past decade, the use of HHE devices and larger laptop size systems to augment physical examination and perform "focused cardiac ultrasound" has become more widespread.[16] International cardiac imaging societies have recommended that focused cardiac ultrasound should not attempt to be comprehensive, but should rather be limited to specific cardiac diagnostic targets, such as the qualitative assessment of ventricular systolic function or the assessment for pericardial effusion.[16,17] In line with these recommendations,

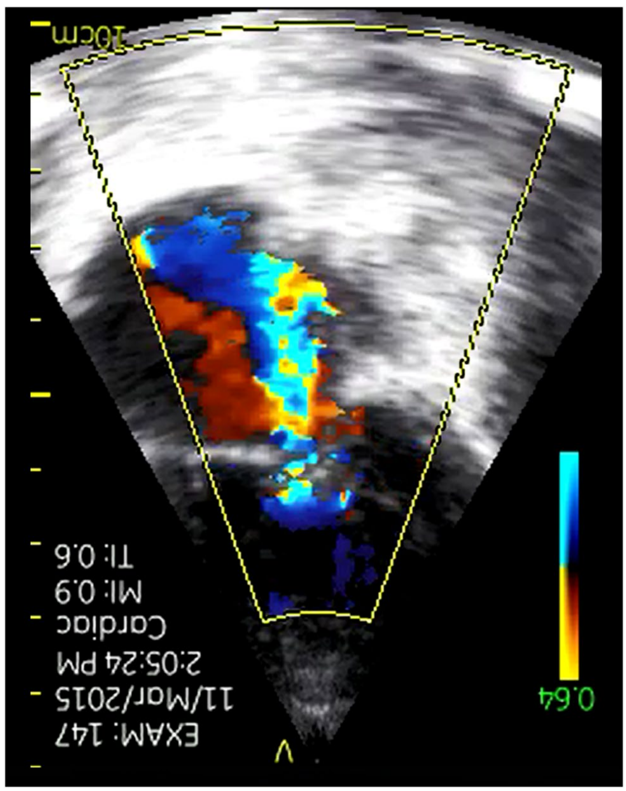

FIGURE 2 HHE color Doppler imaging via pediatric apical view; Subject #2 (10-month old) with moderate tricuspid regurgitation

our HHE protocol focused on right ventricular systolic function and tricuspid regurgitation. Other evaluations such as shunt and aortic arch patency, although clinically important, were not evaluated since we felt that such targets were not within the capabilities of HHE or focused cardiac ultrasound. It seemed more prudent to leave the assessment of targets like these to more traditional methods, eg, SE or physical exam. Maintaining a limited scope of practice is important since, among many other reasons cited in the recommendations, the bedside cardiologist performing HHE is unable to review prior studies for comparison. In our study, HHE performed well in this prescribed, focused role identifying RV systolic dysfunction and higher grade TR in a dichotomous manner, eg, whether or not the RV systolic function is normal or whether or not there is at least moderate TR. This study also demonstrated that HHE could help diagnose RV systolic function deterioration and/or TR progression in individual participants who had more than one encounter during our study period (Table 2).

Intensified outpatient monitoring of HLHS patients, which includes regularly scheduled SE, have resulted in improvement in patient outcomes during the interstage period following the Stage I Norwood procedure.[18,19] The proactive use of bedside focused ultrasound and HHE may be able to identify depressed ventricular function or higher grade atrioventricular regurgitation in HLHS patients before it becomes otherwise clinically evident. Keeping in mind the perils of "incomplete" cardiac evaluations, HHE has a potentially promising role as an adjunct to standard clinical inpatient or outpatient evaluations. During clinical emergencies, HHE may prove invaluable in aiding the bedside clinician to assess for deterioration in RVSF or TR earlier and perhaps identify a cardiac crisis more promptly. HHE may also play a role in predictive modeling and early warning systems, enabling the clinician to have a "quick-look" in premorbid clinical scenarios, thus providing some guidance regarding the need to intervene prior to impending cardiopulmonary arrest.[20] However, identifying the exact etiology of the cardiac crisis would be beyond the scope of HHE and is better ascertained by more standard clinical or cardiac imaging techniques. The use of HHE and focused cardiac ultrasound should be limited to individuals with proven competency. In addition, image storage on shared databases will be critical for communication and quality assurance. How HHE would truly impact outcomes and cost of care in patients with HLHS should be a target of future research, particularly since HLHS continues to have a high associated mortality risk and remains among the most costly diseases in pediatric medicine.[21]

4.1 | Limitations

This was a single-center study during which participants were recruited and HHEs were performed on a convenience basis. Some participants had more than one encounter during the study period and this may have impacted blinding. Interobserver variability likely added to discordance of the evaluations, particularly since the majority of comparisons were based on qualitative assessments of RV systolic function and TR. While RV fractional area change calculation seemed to add some validity to our qualitative 4-point scale to assess systolic function, this measurement seems also to have limited accuracy and interobserver reproducibility in HLHS.[22] MRI is the effective gold standard for assessment of RVSF, but the comparison of HHE to MRI was not feasible since very few patients in the target population (interstage infants) undergo MRI at our institution on a regular basis.[23] Therefore, some concordance maybe inflated due to comparing the same imaging modality on different platforms.

5 | CONCLUSION

As a bedside tool, HHE has the potential to aid in the rapid recognition of depressed ventricular function or significant TR in patients with HLHS. How this technology can be integrated into the clinical care of infants and children with HLHS and other congenital heart defects should be the topic of future research.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest with the contents of this article.

AUTHOR CONTRIBUTIONS

All authors contributed to analysis, drafting, revision, and approval.
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