



# Stress Echocardiography in Pediatric and Adult Congenital Heart Disease: A Complement in Anatomical and Functional Assessment

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**Abstract:** Functional classification of children and adults with repaired and unrepaired congenital heart disease is a challenge for clinicians, due to the heterogeneity of congenital heart disease. Functional studies may be complemented with a stress echocardiogram, which analyzes the hemodynamic behavior of surgical repair zones, residuals, and sequelae. The integration of the anatomical and functional classification criteria developed for congenital heart disease and the results of a stress echocardiogram can establish a more precise functional classification. Stress echocardiograms also provide early diagnosis of functional complications of the congenital heart, allowing timely management decisions. This paper reviews the most important aspects of stress echocardiograms in pediatric and adult congenital heart disease, seeking to spark cardiologists' interest in extending its applications in congenital heart disease. (Curr Probl Cardiol 2021;46:100762.)

## Introduction

**T**he congenital heart disease (CHD) subspecialty has seen great advances over the last few years. Beginning with the development of pediatric cardiology at the start of the 20th century, great strides

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in pediatric cardiovascular surgery and postoperative cardiovascular care followed from the 70s to the 90s.<sup>1</sup> The subspecialty's constant evolution and development has led to the repair of a large number of CHDs, with a greater than 85% pediatric survival rate. As a result, the number of adult survivors of CHD repairs has increased in the last few decades.<sup>2</sup>

The introduction of echocardiography in the 50s was a great help in understanding the anatomy and assessing the hemodynamic function of the heart.<sup>3</sup> For both children and adults with repaired and unrepaired CHD, the introduction of new equipment with greater spatial and temporal resolution, inclusion of harmonics and advanced heart study software provided a greater anatomical and functional understanding of congenital defects, especially complex defects, thus achieving studies which are so exact they are almost equal to the invasive ones.<sup>4</sup>

Stress echocardiograms (SEs) have been well applied in adults, with a special focus on the study of coronary artery disease (CAD). Their application has spread to the study of valve disease, dyspnea, pulmonary hypertension, high-altitude pulmonary edema, and myocardial viability, among others.<sup>5</sup> In the last few years, they have become important in the study of adults with CHD.<sup>6</sup> They have not been used much in the pediatric population, and published studies to date are relatively scarce. The first applications were aimed at studying acquired or Kawasaki syndrome CAD.<sup>7,8</sup> Subsequently, their application was extended to the study of transplanted hearts, congenital coronary artery anomalies (anomalous left coronary artery, intramural coronary arteries, interarterial trajectories), heart surgeries with coronary artery reimplantation (Ross procedure, arterial switch, Nikaidoh procedure), and cancer cardiology (anthracycline toxicity).<sup>9</sup>

This paper aims to raise the awareness of cardiologists dedicated to the care of children and adults with CHD on the importance of systematically and extensively applying SE to the anatomical and functional assessment of CHDs.

## *Anatomical Echocardiographic Assessment*

Classically, the study of CHDs includes the diagnostic sequential segmental analysis proposed by Richard Van Praagh in the 70s.<sup>10,11</sup> This analysis divides the heart into 3 basic areas, sectors, or segments:

1. atrial (or visceratrial) segment
2. ventricular segment
3. arterial segment

The diagnostic process entails a detailed sequential morphologic analysis to determine the atrial situs, abdominal situs, right or leftward orientation, and morphology of the structures. Subsequently, the type and mode of connection between the various cardiac segments is analyzed, complemented by a functional hemodynamic analysis in keeping with the CHD found.

In general, the order includes:

1. defining the atrial situs
  - a. solitus
  - b. inversus
  - c. isomeric (dextromorphism, levomorphism)
2. defining and analyzing the atrio-ventricular connection
  - a. type of connection (concordant, discordant, ambiguous, double ventricular inlet, absent connection)
  - b. connection mode (permeable or perforated, nonperforated, straddling, and common)
3. defining and analyzing the ventricular-arterial connection
  - a. type of connection (concordant, discordant, double outlet, and single outlet)
  - b. connection mode (perforated, nonperforated, and straddling)
4. describing the associated defects
5. determining any additional features

Pediatric congenital cardiovascular studies explore CHD in its pure or native form (without repair or intervention) as well as repaired CHD, generally with a very good echocardiographic window. The extent of functional analysis is basically focused on evaluating the ventricles, the valves present, pulmonary pressure and details related to the surgical repair or intervention. In adolescents and adults, the CHDs have already been operated on in most cases. On occasion, a large number of these patients have undergone up to 2 or 3 prior surgeries in childhood, causing intra- and extrathoracic adhesions and surgical scars which interfere with the echocardiographic window.

Basic and advanced hemodynamic assessment is hampered by the anatomical layout of hearts which have been “transformed by surgery.” Surgical and survival successes have turned the pediatric patient with CHD into an adult postoperative CHD patient. The adult’s postoperative heart is not a healthy heart; it is a transformed, “neo-formed,” heart. Its original, or native, anatomy has been modified with synthetic CHD repair elements (patches, valves, intra- or extracardiac tubes, stents) which modify not only its primary anatomy but its whole physiology.<sup>12</sup>

Sixty percent of CHDs are known to be diagnosed and treated in the first year of life, and 30% throughout childhood and adolescence up to 16 years of age. It is estimated that 10% are diagnosed in adulthood.<sup>13</sup> In this scenario, we have 2 types of patients (repaired and unrepaired); at the same time, the repaired cases may have incomplete treatment or partial repairs for various reasons (eg, the physiologically single ventricle heart which did not reach the Fontan stage, remaining at the Glenn stage).

From my expert point of view, I recommend that the diagnostic sequential segmental study guidelines used in pediatrics be extended and applied in the same way in adults, in order to maintain the correct anatomical and functional assessment of CHDs.

For CHDs which have been surgically repaired, 3 questions should be answered with the initial echocardiographic assessment.<sup>14</sup>

1. Is there residual CHD?
2. What hemodynamic sequelae does or will it have?
3. What complications does or could it have?

1. Residual: “remains, what is left.” These are lesions deliberately left at the time of CHD repair. With few exceptions, they are obligatory defects (small residual ventricular septal defects [VSDs], left atrioventricular valve clefts following complete AV canal repair; obligatory atrial septal defects in the case of CHD repair with pulmonary hypertension).

2. Sequelae: What “arises or follows” from a prior event; these are new cardiovascular disorders produced after carrying out CHD repair. They are lesions which are inherent to the repair, and are represented by disorders intentionally incurred during surgery (pulmonary regurgitation following tetralogy of Fallot repair).

3. Complications: Undesired cardiovascular or systemic disorders resulting from the therapeutic procedures, or which appear during the natural evolution of the CHD (complete AV block, infectious endocarditis and ictus, among others).

## *Functional Class Assessment*

For adult and pediatric patients, the functional class (FC) assessment is based on the classic New York Heart Association classification and Ross classification,<sup>15,16</sup> respectively. These may not be deliberately used in CHD patients, particularly in complex cases and even less in unrepaired cases. Many of these patients have been ill for years and have never really experienced a normal FC. Clinical deterioration symptoms are often minimized and masked in both adolescents and adults.

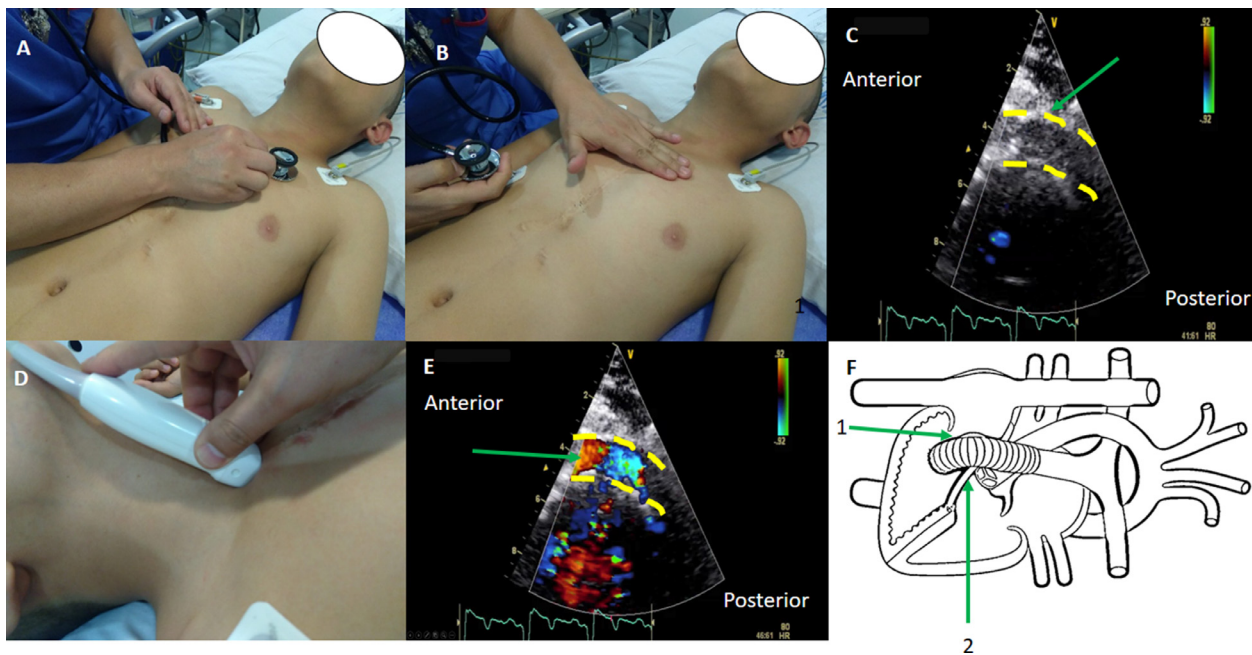
For this population, it is more appropriate to apply the anatomic and physiological classification (APC) which integrates the anatomical or morphologic part of the repaired or unrepaired CHD with the New York Heart Association FC and 9 clinical variables (hypoxemia, pulmonary hypertension/pulmonary arterial hypertension, hemodynamically significant shunt, venous and arterial stenosis, exercise capacity, end-organ dysfunction, concomitant acquired valve disease, arrhythmia, and aortopathy) which, if present, add severity to the CHD. The classification establishes 4 physiological stages in order of severity (A, B, C, and D).<sup>17</sup> In order to achieve a more precise FC in this population, my expert recommendation is to also extend it to the adolescent population.<sup>18</sup>

In expert hands, pediatric and adult echocardiography achieves a very good, highly precise anatomical assessment in most cases, as well as a sufficient functional assessment to allow clinical cardiologists and surgeons to make management decisions.<sup>19</sup> However, in adolescents and adults, image resolution is not the best, especially in complex CHDs. Consequently, the precision of the anatomical and functional assessment is limited. The echocardiographic windows commonly used in adults with noncongenital hearts are altered by the CHD itself, as well as its surgical repair. The constant evolution of repair residuals and sequelae distort the normal architecture of the heart. Therefore, the intra- and extracardiac structures are often not in their usual positions. In these cases, the cardiologist performing the echocardiogram must be very familiar with the original CHD, as well as the surgical technique used for repair. Having these points clear, variations of the classical echocardiographic windows may be employed for a better anatomical and functional exploration<sup>20</sup> (Fig 1).

The conventional treadmill stress test (TST) is a good complement in the functional assessment and stratification of CHD; the values obtained in response to exercise should be correlated with the symptoms experienced. The interpretation should always be individualized, taking into account the type of CHD being evaluated and whether there are residual lesions or sequelae.<sup>21</sup> With the results of this study and the application of the APC criteria, a more precise stratification may be achieved.

In general, a conventional stress test allows us to evaluate:

- a. hemodynamic parameters: chronotropic response, pressor response, double product (systolic pressure x frequency)
- b. electrocardiographic parameters: ST elevation or depression, arrhythmias, or conduction disorders
- c. functional capacity: the capacity for exertion of the patient and his/her heart. It is measured in METs (the amount of oxygen consumed



**FIG 1.** Young adult repaired of double outlet RV, with Rastelli technique. Echocardiographic study is guided by the signs found in the physical examination, which correspond to the projection of the extracardiac structures (extracardiac tube). (A) left infraclavicular murmur; (B) fremitus in the same localitation; (C) green arrow shows the projection of the extracardiac tube (just where is fremitus); (D) unconventional suprasternal window, to explore extracardiac and anterior tube; (E) Doppler color through extracardiac tube (green arrow); (F) Rastelli surgery scheme (shows tube in extracardiac and anterior projection); (1) extracardiac tube connecting RV with pulmonary artery; (2) closed ventricular septal defect with patch.

by an average individual at rest). In a TST using the Bruce protocol, the number of METs is usually equal to the number of minutes walked on the treadmill plus one.

Conventional TST neither show the heart's dynamics during exercise nor how its valves and other related structures function. When this study is performed on a CHD patient, it is important to observe the hemodynamic response of these structures as a function of the CHD, with a special focus on assessing the surgical repair zones.

Surgical repair zones, those which contain synthetic material (Gore-Tex patches, pericardium, and defect closure devices such as Amplatzer, among others) (Fig 2), are non-natural areas of the heart. The synthetic materials required for surgical repair do not have the biological behavior of heart tissue (the ability to contract, relax, and distend); therefore, it is expected that they will not contribute to cardiac output or ventricular mechanics during heart stress. The greater the number of foreign elements, the lower the normal heart response to stress.<sup>22</sup>

### *Stress Echocardiography and Implications Related to CHD*

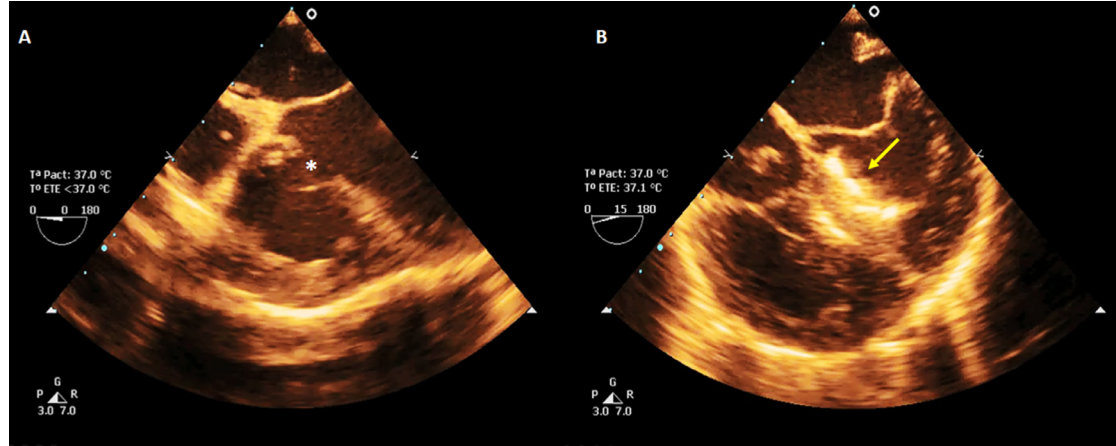
Stress echocardiography (SE) allows a better hemodynamic and functional assessment of congenital hearts in both children and adults. The functional information which can be obtained by studying the stressed heart allows a correlation of the CHD findings with the residuals and sequelae present and the type of surgical or interventional repair employed.

There are currently several indications for SE in CHD, but the intention is to extend the application to more repaired and unrepaired CHDs, which would assist clinical decision-making in many cases, without treatment delays.<sup>23</sup>

### *General Recommendations for SEs in CHD*

Taking into consideration the recommendations for stress echocardiography in adults without CHD,<sup>24</sup> the following recommendations may be added for repaired and unrepaired CHDs:

- know the underlying CHD very well.
- have a clear understanding of the surgical history, especially the type of intervention, date, surgical repair materials and technique employed.
- perform a very complete and detailed anatomical study of the CHD prior to the test, paying special attention to the surgical repair zones.



**FIG 2.** (A) Transesophageal apical 4-chamber view shows large VSD (\*) in a child; (B) the same in A, but closed VSD with Amplatzer device interventional repair zone, which contain synthetic material (yellow arrow shows Amplatzer device), do not have the biological behavior of heart tissue.



Describe the residual lesions (small septal defects, valve or outflow tract stenosis, among others), sequelae (valvular regurgitation, and atrial and ventricular dilatation, among others) and complications present. This type of study should, if possible, be performed by the same cardiologist who will be performing the SE.

- be aware of the patient's medications which may affect the measurement parameters (beta blockers, diuretics and antiarrhythmics, among others).
- note the presence of devices (pacemakers, defibrillators).
- the use of harmonic imaging minimizes near field artifacts, improves resolution, intensifies myocardial signs and is better than fundamental imaging for viewing the endocardial border.
- the use of contrast is recommended when two or more segments are not visualized well.
- exercise stress is recommended for all patients able to exercise, since the ability to exercise is an important predictor of results. Either treadmill or stationary bicycle exercise may be used for the test. It is recommended that exercise be limited by the patient's symptoms, within a standardized protocol in which the workload is gradually increased by stages.
- the Bruce protocol is most frequently used for treadmill exercise SEs.
- images are obtained at rest and immediately after finishing the exercise.
- bicycle SE may be performed in either erect or supine position; one advantage is that the image may be obtained during exercise. With the usual supine bicycle protocol, a baseline image is obtained, with subsequent images after an initial workload of 25 W, at stress peak and during the recovery phase. The workload is increased by 25 W every two or three minutes.
- pharmacologic stress is recommended for patients who cannot exercise due to any condition.
- the most frequently used medication is dobutamine, beginning with a progressive infusion of 5 micrograms per kilo per minute (mcg/kg/min), increasing at 3-minute intervals to 10, 20, 30, and 40 mcg/kg/min. It has an advantage over other medications in assessing regional wall motion.
- vasodilators are an alternative. Their advantage over dobutamine is their usefulness in evaluating myocardial perfusion. The inclusion of low-dose stages allows viability and ischemia to be identified in

segments with abnormal function at rest, even if the viability assessment is not the main objective of the study.

- the objective of SE is best achieved when the target heart rate is reached (85% of the maximum heart rate for age). It is designed to find new abnormalities in wall motion or worsening of segments with baseline alterations, significant arrhythmias, hypotension, severe hypertension or angina symptoms.

In some CHDs, chronotropic incompetence is common, due to palliative or repair techniques or procedures carried out in childhood. Neonatal balloon atrial septostomy for transposition of the great arteries is associated with sinus nodal artery damage and the development of sinus node dysfunction in the second decade of life. The Fontan procedure is another common cause of sinus node dysfunction in the adult.<sup>25</sup>

Atropine may be used, at 0.25-0.5 mg up to a total of 2 mg, to inhibit vagal tone and increase the heart rate when this is not achieved with the maximum dose of dobutamine. It is not often used in children; if needed, the recommended dose is 0.01 mg/kg, with a maximum of 0.6 mg per dose.

### *Principal congenital heart disease in which stress echocardiograms are used, and to which they could be extended*

In 2017, the European Association of Cardiovascular Imaging and the American Society of Echocardiography published “The Clinical use of SE in Non-Ischaemic Heart Disease.” Among these diseases are various CHDs<sup>6</sup>:

**Atrial Septal Defect.** Simple, small defects which are closed early in life do not have any repercussions in adults. Large defects which are closed late leave sequelae in heart rhythm and function. Those related to the superior sinus venosus may be complicated by sinus node damage and dysfunction.<sup>26</sup>

Stress echocardiography can evaluate RV function; the most common methods employed are TAPSE and fractional area change (FAC), and pulmonary pressure at rest and during exercise, which is an indicator of increased pulmonary vascular resistance.<sup>27</sup>

**Ventricular septal defect.** The ventricular septum plays a large role in ventricular mechanics; it is responsible for achieving shortening and decreased left ventricular (LV) area. It is very clear that when the septum

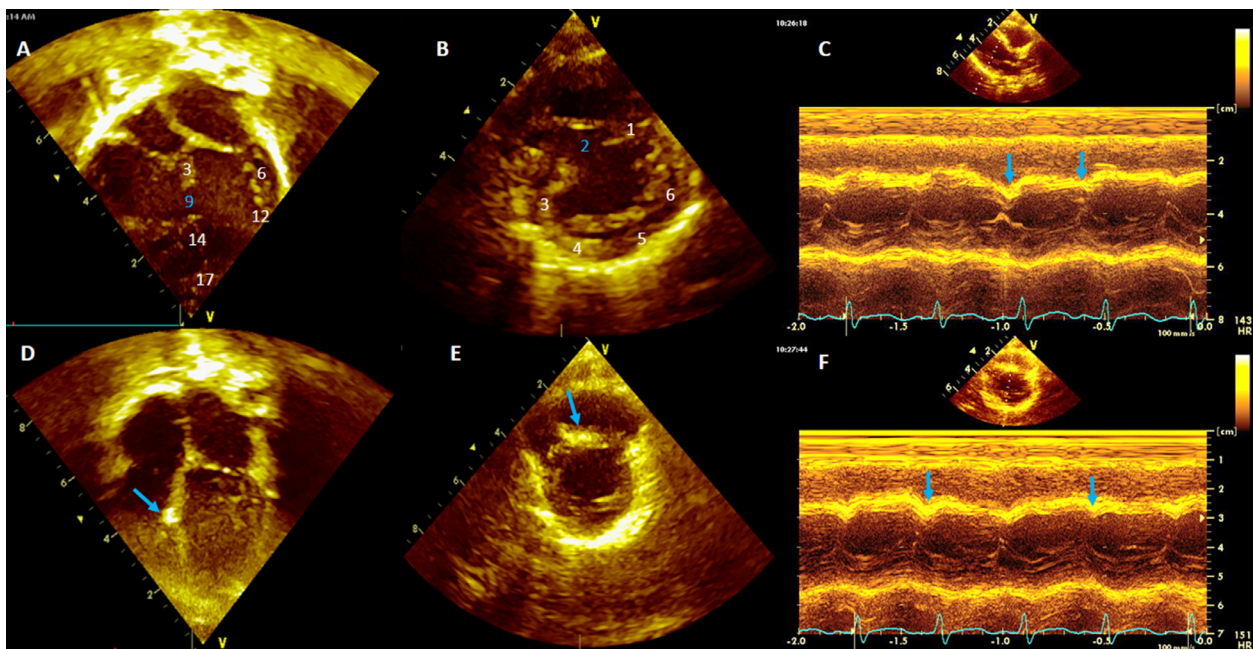
has ischemic zones, LV function is compromised to varying degrees. VSDs may be located at different levels (membranous, muscular, inlet, or outlet) and have different sizes. The 17-segment model proposed for segmental evaluation of the LV should be applied with caution in these cases; these hearts were born without a portion or segment (Fig 3) and ventricular mechanics are not expected to be the same as in a heart without a congenital septal defect. Large VSDs already have altered ventricular mechanics and function in infancy; even after very good surgical repair, the mechanics will continue to be abnormal. This abnormality will become more obvious and be more significant in adulthood with the acquired adult diseases (arterial hypertension, dyslipidemia, diabetes, obesity, and CAD, among others) which add cardiovascular risk factors to a congenital heart with preexisting underlying functional alterations.

For years, it was thought that small VSDs which did not close naturally and were not closed through surgery or interventional procedures were benign. However, it has been shown that these VSDs become complicated in the third or fourth decade of life. Complications such as arrhythmias, double-chambered right ventricle, aortic regurgitation and infective endocarditis have been described.<sup>28</sup>

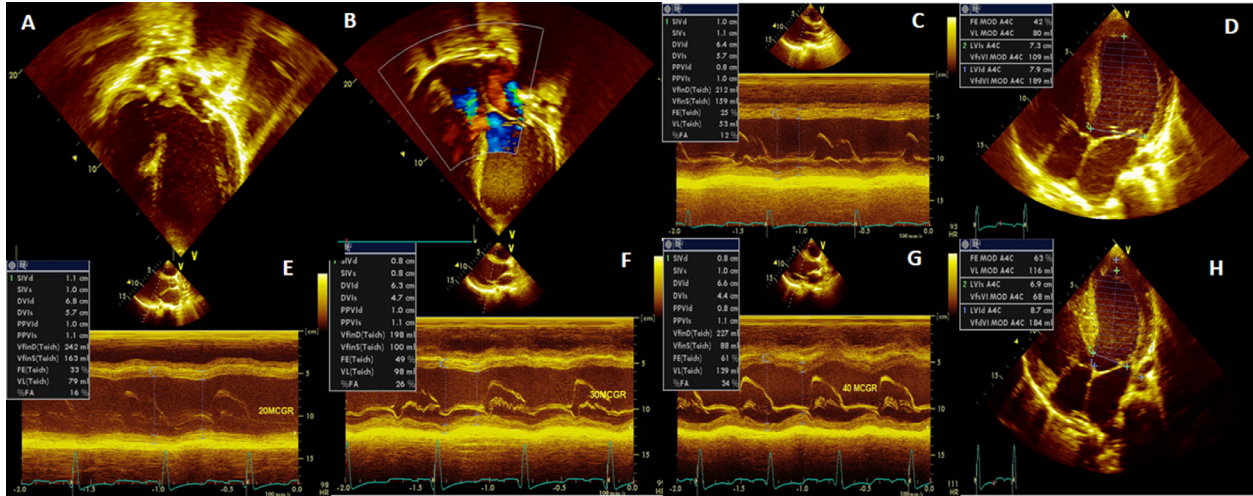
The VSD repair zone is an inert tissue without the ability to contract or shorten; the larger it is, the more it compromises the mechanics of ventricular contraction. During ventricular septal analysis, it is recommended that the degree of shortening, degree of mobility and the effect of the natural septal zone on function (before and after the closed defect) be described (Fig 4). In addition, peak systolic and diastolic velocities ( $s'$  and  $e'$ ) can be measured at rest and during exercise. The cardiologist should closely observe the movement of the repaired zone compared to the unrepaired zone, as well as detect the presence of arrhythmias arising from the surgical repair zone or new stress-induced hemodynamic alterations.

When assessing an unrepaired VSD, changes may be found in the gradients and flow patterns which may be clinically significant. This is the case with VSDs which, during maximal effort, may reverse the flow direction (from left-to-right at rest, to right-to-left at maximum effort) due pressure changes in the ventricles. This causes decreased arterial oxygen saturation and fatigue. Subaortic VSDs may cause trivial aortic valve regurgitation which may increase during effort. Also, scar tissue next to the VSD may cause dynamic obstruction gradients with effort.

**Univentricular Heart.** A few studies have been done with a small number of pediatric patients who have undergone the Fontan procedure. These have used the supine exercise SE modality, analyzing caval blood



**FIG 3.** (A) Apical 4-chamber view shows large unrepaired muscular VSD in a 6-month-old child. In relation to the cardiac segments in this projection, segment 9 is absent (corresponds to the muscular VSD); (B) short basal axis view shows segment 2 is absent (corresponds to the muscular VSD); (C) analysis M mode LV long axis view in the same patient, 6 months after surgical repair shows abnormal septal contraction in repair area (blue arrow) under repose or baseline conditions; (D) apical 4-chamber view shows VSD closure patch; (E) short basal axis view shows VSD closure patch; (F) analysis M mode LV short basal axis view shows abnormal septal contraction in repair area (blue arrow) under repose or baseline conditions.



**FIG 4.** This case shows systolic LV dysfunction in a 5-year-old child with unrepaired VDS. Stress echocardiography study showed that LV function improved in response to dobutamine. Then surgical VSD repair was performed. (A) Apical 4-chamber view shows unrepaired large inlet VSD in a child; (B) the same in A, Doppler color with left to right shunt and dilated left ventricle (LV); (C) under baseline conditions, M mode LV long axis view shows ejection fraction (EF): 25%; (D) apical 4-chamber view, EF: 42% Simpson method; (E) dobutamine stress echocardiography with 20 mcg/kg/min infusion, M mode LV long axis view shows increased EF: 33%; (F) same projection in E, with increasing dobutamine infusion 30 mcg/kg/min, M mode LV long axis view shows increased EF: 49%; (G) same projection in E, with increasing dobutamine infusion 40 mcg/kg/min, M mode LV long axis view shows increased EF: 61%; (H) apical 4-chamber view, EF: 63% Simpson method, with dobutamine infusion 40 mcg/kg/min.

flow at rest and during exercise; however, they used cardiac resonance,<sup>29</sup> which is expensive and impractical.

Interpretation is more difficult in univentricular hearts due to the great heterogeneity of CHDs. Heart defects which have been taken to univentricular circulation with the Fontan procedure, regardless of the baseline heart disease, are hearts with a single functional systemic ventricular chamber, which could have left, right, mixed or indeterminate morphology. Whatever its form, the 17-segment motion indices cannot be applied.

The analysis aims to measure qualitative variables such as degree of motion, degree of shortening, wall synergy, and the detection of hypokinetic and akinetic zones. It also measures reproducible quantitative variables like MAPSE, TAPSE, FAC, and chamber dilatation, among others. The changes in the pattern of diastolic function at rest and during maximum exercise can be quantified by Doppler; the appearance of various degrees of atrioventricular valve regurgitation and atrial dilatation can be easily measured. The measurement of peak systolic and diastolic velocities ( $s'$  and  $e'$ ) in the free walls is also recommended. When there is extracardiac tube fenestration, changes in flow direction (venous-arterial, bidirectional, or reversed) during exercise can be easily analyzed, and reflect the hemodynamic response of systemic and pulmonary pressures to exercise.

Finally, the outlet flow patterns of the univentricular heart should be analyzed. The quantitative means (outflow tract gradient, velocity-time integral, stroke volume and cardiac output) should be determined and compared at rest and during exercise.

***Repaired Coarctation of the Aorta.*** A common complication for children and adults is re-coarctation, which may have a low intensity at rest and increase significantly during exercise. There is an increased risk of systemic arterial hypertension in survivors. Using suprasternal views, SE is useful in measuring gradients at rest and during exercise, along with a complete qualitative and quantitative functional assessment of the LV.<sup>30</sup> Significant gradients during exercise (mean gradient >30 mm Hg at any stage) together with systemic arterial hypertension are very suggestive of significant coarctation. The hemodynamic analysis becomes more interesting when it is extended to the associated defects. Bicuspid aortic valve is known to be commonly associated in 85% of cases, and can be accompanied by subvalvular, valvular, or supravalvular aortic stenosis and malformation of the mitral valve.<sup>31,32</sup> The hemodynamic analysis should include all possible variables which can be measured and quantified.

**Other Congenital Defects of the Aortic Arch.** Stress echocardiography can also be applied to interrupted aortic arch, pseudocoarctation, double aortic arch, vascular rings, and truncus arteriosus. The objective is to reveal subclinical obstructions which may be treated early.

**Tetralogy of Fallot.** Being a common CHD in children, it is one of the most prevalent CHDs in adults. The different surgical repair techniques leave residuals and sequelae (pulmonary regurgitation [PR], VSD, residual RV outflow tract obstruction, and pulmonary branch stenosis, among others) in various heart zones (ventricular septum, RV outflow tract, pulmonary valve, pulmonary trunk and branches), which negatively affect RV function. Several studies applying SE have been carried out in pediatric and adult patients, analyzing the functional parameters of the RV (FAC, TAPSE, synchrony indices), which have shown a very good correlation between the results and lesions found.<sup>33-36</sup>

After extensive surgeries in childhood which tend to include infundibular resection, right ventriculotomy in some cases, muscle band resection, pulmonary valve resection and pulmonary trunk reconstruction, abnormal RV mechanics are to be expected. The main sequelae include RV outflow aneurysms, RV dilatation, and PR. Among the most frequent residuals are VSD, RV outflow obstructions, and pulmonary branch stenoses. Aortic root dilation and secondary aortic regurgitation is a long-term complication.<sup>37</sup> In addition, ventricular and supraventricular arrhythmias develop. Finally, RV failure is the result.

Stress echocardiography analyzes RV radial contractility with special attention to the infundibular region, longitudinal free wall contraction measured by TAPSE and global contractility quantified by FAC. This is followed by a Doppler pattern analysis of the RV outflow tract, the pulmonary valve (if possible, extending to the pulmonary branches), and peak systolic and diastolic velocities ( $s'$  and  $e'$ ). This is coupled with the Doppler pattern of the tricuspid valve, measuring the degree of regurgitation, correlated with the size of the right atrium.

**Systemic Right Ventricle.** Functional evaluation of a systemic RV (SRV) is a great challenge for all cardiologists. These conditions are estimated to account for 10%-12% of all CHDs. Among the most frequent are congenitally corrected transposition of the great arteries, transposition of the great arteries repaired with atrial switch, double inlet RV, hypoplastic left heart syndrome, and unbalanced atrioventricular canal (undergoing Fontan procedure).<sup>38</sup>



In most cases, SRV begins to show signs of dysfunction in the second decade of life and often requires heart transplantation between the third and fourth decades, due to SRV failure.

It is recommended that, for TCCGA, SE should take baseline diastolic measurements of the ventricles, FAC, SRV ejection fraction, and systemic TAPSE. The Doppler parameters should include measurement of the degree of regurgitation of the systemic-tricuspid valve, and peak systolic and diastolic velocities ( $s'$  and  $e'$ ) taken from the free wall. Almost 80% of cases are associated with outflow tract obstructions; the most common cause is a naturally closing VSD which causes dynamic obstruction of the outflow tracts (subpulmonary or venous SRV or LV). This obstruction can be measured at rest and during exercise. In transposition of the great arteries after atrial switch, one of the main problems is chronotropic incompetence due to sinus node dysfunction, which may be seen in the second decade of life. Other complications include intra-atrial reentry tachycardia, atrial baffle obstruction or leak and pulmonary arterial hypertension. Under normal conditions, the atria are able to distend and increase their capacitance. In this case, the neo-atria are rigid due to the elements used to create the baffles; thus, they are unable to dilate and increase their capacity during intense exercise. Stress echocardiography must be very well selected and applied with clinical discretion due to the high risk of decompensation in this group of patients.

For all other CHDs with SRV which have undergone the Fontan procedure, the study will focus on the points previously described in the section on univentricular hearts.

**Complete Atrioventricular Canal.** This CHD is common in children. Repair usually includes closure of the septal defects (atrial and ventricular) with a single- or double-patch technique, completing the procedure with atrioventricular valve repair and cleft closure. The repair must be very precise in order to preserve the function of the atrioventricular valves. If there are no immediate complications after surgery (atrioventricular block), survival will be similar to that of the general population. Following septal defect closure, the mechanics of contraction will understandably never be normal, and the SE should include the portion of the inflow septum repaired with a patch. In addition, the diastolic pattern of the atrioventricular valves may be altered since the left atrioventricular valve now has 3 leaflets, unlike the normal mitral valve which only has 2. This explains why diastolic dysfunction during exercise may occur sooner or later, more often in cases with a residual cleft.



Ventricular qualitative variables are assessed, with special attention given to the diagnosis of hypokinesias or akinesias in the repair zones, as well as wall motion and thickening.

These variables are complemented with a quantitative analysis such as measurement of ventricular inflow and outflow gradients, time velocity integral means, and stroke volume. Pulmonary pressure, diastolic function, and peak systolic and diastolic velocities ( $s'$  and  $e'$ ) of the ventricular free wall should be analyzed.

**Anomalous Pulmonary Venous Return.** Final repair includes the connection of the left atrial collector to either the posterior wall or roof, usually through a wide anastomosis. In some cases, the progression of scarring may cause obstruction of the anastomosis and create significant stenosis leading to postcapillary pulmonary hypertension and even pulmonary edema.

The diastolic pattern of the collector should be analyzed at rest and compared to that during exercise. According to the pulmonary dynamics and relationship of the collector to the mediastinal structures, there may be an increased gradient causing dyspnea on exertion. The results are correlated with the symptoms and the presence of pulmonary hypertension.<sup>39,40</sup> All of this accompanied by the complete RV and LV function.

## Conclusions

Stress echocardiography provides a complementary functional assessment of repaired and unrepaired CHDs, integrating anatomical findings related to residuals and sequelae and their hemodynamic response to stress. This, together with the clinical manifestations and APC, allows a more precise functional stratification. As its application spreads to various CHDs, it allows us to be aware of signs of decompensation earlier, so as not to delay corrective measures. Applying the CHD assessment and interpretation recommendations, SE may be safely performed using exercise or drug stimulation.

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