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Commentary on the New 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease

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Introduction

The constant growth in the population of adults with congenital heart disease (ACHD) has taken many countries by surprise. Canada is a pioneer and world leader in development and care. It was the first country to develop clinical practice guidelines for ACHD in 1996, followed by the United States in 2008 and Europe in 2010 [1]. In recent years, a few other consensuses have been developed along with scientific statements in America and Europe, which have been adapted to the needs of this growing population. These serve as tools for managing ACHD complications and relapses which are periodically discovered as more children survive multiple and innovative surgical procedures [2-5]. The most recently published guidelines [6] constitute an update developed in collaboration with the American Association for Thoracic Surgery, the American Society of Echocardiography, the Heart Rhythm Society, the International Society for Adult Congenital Heart Disease, the Society for Cardiovascular Angiography and Interventions, and the Society of Thoracic Surgeons. World-renowned experts on the writing committee and members of the ACC/AHA task force have provided a 171 page document, 110 pages of which are entirely devoted to recommendations, with 878 references. The guideline contains new and interesting recommendations which have evolved and seek to parallel the growing number of ACHD survivors, most of whom have medium and high complexity

defects. They are clearly applied to individuals over the age of 18, and inherited disorders that may have cardiac manifestations, such as Marfan syndrome or hypertrophic cardiomyopathy, are excluded. Also excluded are anatomic variants such as patent foramen ovale, as well as acquired valve disease. Class of recommendation are:

I-benefit >>> risk, with a strong recommendation

IIa-benefit >> risk with a moderate recommendation

IIb-benefit ≥ risk, with a weak recommendation. This is the same as the 2008 guidelines [7].

Class III is now classified as benefit = risk, moderate recommendation, there is no benefit. This new version includes an additional class of recommendation III- (harm) strong, risk > benefit, with a strong recommendation that emphasizes that the interventions clearly do more harm and should be avoided.

Discussion

1. Understanding the heterogeneity of congenital heart diseases and breaking the paradigm of cure

My impartial and critical opinion is that the first paradigm which should be eliminated is that of congenital heart disease (CHD) cure, at least in the vast majority of cases, with the exception of a few simple defects which are considered to be cured. Most CHDs are repaired, substantially improving the original anatomical and functional condition, which in many cases is incompatible with life (hypoplastic left ventricle, pulmonary atresia with intact septum, hypoplastic right ventricle, among others), and transforming that incompatible situation into an almost normal physiology. This teaches us that heart surgery is reparative and not curative. The panel of experts in this new guideline emphasizes this point: "*patients are not cured of their disease following a successful treatment in childhood*". *"Almost all patients will have sequelae of native CHD or surgical repair or palliation, although these sequelae may take decades to manifest themselves*". The great heterogeneity of CHDs is highlighted, explaining why the same CHD manifests in a different fashion between one adult and another. In other words, although it has the same embryological origin, the severity of the same defect varies from case to case. The most typical example is Tetralogy of Fallot; the greater the obstruction and hypoplasia of the pulmonary branches, the greater the morbidity in childhood, requiring prior palliative procedures (Blalock-Taussig systemic-pulmonary fistula) before being repaired. Likewise, the residuals, sequelae and complications will be greater compared to those with better anatomy who did not go through those prior palliations. This heterogeneity and the long symptom-free intervals limit the ability to generate data applicable to the whole ACHD population, or for adults with specific lesions or repairs. As a recommendation, a CHD should never be discharged from the cardiology department: care and follow up should be lifelong.

2. From the anatomical classification to the anatomic-functional classification

This is a big change from the 2008 guidelines, and none of the clinical practice guidelines for ACHD from Europe or North America had ever tried to classify ACHD as a functional and anatomical state, at the same

time. Beginning with the Bethesda conference in 2001 [8], all the previous guidelines had taken purely anatomical data as a reference for both repaired and unrepaired CHD, to establish complexity. However, as explained in the previous point, ACHDs are highly heterogeneous, and this means that severity should be classified not just based on native anatomy, but also according to comorbidities and complications which have occurred throughout the evolution of the congenital defect, correlating all of this with the current functional class. For many authors, the New York Heart Association (NYHA) functional parameters for establishing functional class are not exact when applied to CHD, since these assessment recommendations have been developed for non-congenital hearts [9]. However, the functional classification system continues to be useful, and is included in the current guidelines.

The anatomic-physiological (AP) classification includes nine clinical variables and the NYHA functional classification. The clinical variables are:

2.1 Hypoxemia/Hypoxia/Cyanosis: Hypoxemia is defined as oxygen saturation measured by pulse oximetry at rest ≤90%. It is considered to be severe when the oxygen saturation at rest <85%.

The clinical sign of hypoxemia is cyanosis: a bluish tinge to the skin and mucous membranes which appears when the reduced capillary hemoglobin is > 5g/dl. Its appearance depends on the amount of hemoglobin present. An anemic patient must have a greater percentage of desaturation in order to achieve 5 g/dl of reduced hemoglobin and show cyanosis, while cyanosis will be more intense with increased hemoglobin levels [10].

Cyanosis may be:

- **peripheral:** caused by increased oxygen extraction in the tissues, or

- **central:** caused by decreased oxygen saturation in arterial blood.

Hypoxemia is a central cyanosis (decreased oxygen saturation in arterial blood), caused by various cardiovascular disorders with a right to left shunt, leading to mixing of venous or unsaturated blood with arterial blood.

2. 2. Pulmonary Hypertension (PH) / Pulmonary Arterial Hypertension (PAH):

PH is defined as mean pulmonary pressure (PM) by right heart catheterization ≥25mm Hg.

PAH is defined as PM by right heart catheterization ≥25mm Hg and pulmonary capillary wedge pressure ≤15 mm Hg and pulmonary vascular resistance ≥3 Wood units.

2.3. Hemodynamically significant shunt: an intracardiac shunt is hemodynamically significant if there is evidence of chamber enlargement distal to the shunt and/or evidence of sustained $Q_p : Q_s \geq 1.5:1$.

An intracardiac shunt not meeting these criteria would be described as small or trivial.

2.4. Venous and arterial stenosis: may occur at several levels such as

- aortic recoarctation
- supravalvular aortic obstruction
- venous baffle obstruction
- supravalvular pulmonary stenosis
- branch pulmonary artery stenosis
- cavopulmonary connection stenosis
- pulmonary vein stenosis

2.5. Exercise Capacity: patients with CHD are often asymptomatic, notwithstanding demonstrated exercise limitations on objective evaluation. Both subjective and objective exercise capacity assessment is important, as this is associated with the prognosis.

2.6. End-organ dysfunction: clinical and/or laboratory evidence of end-organ dysfunction (kidney, hepatic, liver, lung).

2.7. Concomitant acquired valve disease: classified as mild, moderate or severe

2.8. Arrhythmia: This point is very important becasuse arrhythmias make up 90% of the consults and hospitalization. By frequency, they are atrial fibrillation and flutter in 80% of cases, ventricular tachycardia (VT) 5-7%, symptomatic bradycardia 3-5%, and pacemaker malfunction 5%. My impartial and critical opinion is they are underestimated so many times, they increase the risk of sudden death, especially in complex CHD. The causes are multifactorial and occur in both repaired and unrepaired CHD [11].

- no arrhythmia

- arrhythmia not requiring treatment: bradyarrhythmia, atrial or ventricular tachyarrhythmia not requiring antiarrhythmic therapy, cardioversion, or ablation

- arrhythmia controlled with therapy: bradyarrhythmia requiring pacemaker implantation; atrial or ventricular tachyarrhythmia requiring antiarrhythmic therapy, cardioversion, or ablation; atrial fibrillation and controlled ventricular response; patients with an implantable cardioverter/defibrillator

- refractory arrhythmias: atrial or ventricular tachyarrhythmias currently unresponsive or refractory to antiarrhythmic therapy or ablation.

2.9. Aortopathy: aortic enlargement is common in some types of CHD and after some repairs.
Aortic enlargement may be progressive over a lifetime. There is no universally accepted threshold for repair, nor is the role of indexing to body size clearly defined in adults, as in pediatric populations.

- mild aortic enlargement is defined as a maximum diameter 3.5-3.9 cm.

- moderate aortic enlargement is defined as a maximum diameter 4.0-4.9 cm.

- severe aortic enlargement is defined as a maximum diameter ≥ 5.0 cm.

2.10. NYHA functional classification system:

I- Patients with heart disease with no resulting limitation of physical activity. Ordinary physical activity does not cause undue fatigue, palpitations, dyspnea, or anginal pain.

II- Patients with heart disease resulting in slight limitation of physical activity. They are comfortable at rest, but ordinary physical activity results in fatigue, palpitations, dyspnea, or anginal pain.

III- Patients with heart disease resulting in marked limitation of physical activity. They are comfortable at rest, but less than ordinary activity causes fatigue, palpitations, dyspnea, or anginal pain.

IV- Patients with heart disease resulting in the inability to carry on any physical activity without discomfort. Symptoms of heart failure (HF) or the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort increases.

3. Physiological Stage Classification

A

□ NYHA functional class (FC) I symptoms

□ No hemodynamic or anatomic sequelae

□ No arrhythmias

□ Normal exercise capacity

□ Normal renal/Hepatic/Pulmonary Function

B

□ NYHA FC II symptoms

□ Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction)

□ Mild valvular disease

□ Trivial or small shunt (not hemodynamically significant)

□ Arrhythmia not requiring treatment

□ Abnormal objective cardiac limitation to exercise

C

- NYHA FC III symptoms
- Significant (moderate or greater) valvular disease; moderate or greater ventricular dysfunction (systemic, pulmonary, or both)
- Moderate aortic enlargement
- Venous or arterial stenosis
- Moderate aortic enlargement
- Venous or arterial stenosis
- Mild or moderate hypoxemia/cyanosis
- Hemodynamically significant shunt
- Arrhythmias controlled with treatment
- Pulmonary hypertension (less than severe)
- End-organ dysfunction responsive to therapy

D

- NYHA FC IV symptoms
- Severe aortic enlargement
- Arrhythmias refractory to treatment
- Severe hypoxemia (almost always associated with cyanosis)
- Severe pulmonary hypertension
- Eisenmenger syndrome
- Refractory end-organ dysfunction

Patients should be classified according to the highest relevant anatomical or physiological criteria. The anatomical classification has not changed from the previous 2008 classification, which is:

I: simple (native disease and repaired conditions)

II: moderate complexity (repaired or unrepaired conditions)

III: great complexity (or complex).

The functional class may vary at any moment with the performance of heart disease interventions. For example, an adult with partial or total anomalous pulmonary venous connection (moderate complexity II) may have NYHA FC III symptoms: mild or moderate hypoxemia/cyanosis, a hemodynamically significant shunt, and pulmonary hypertension (less than severe), placing him/her in anatomical-physiological stage IIC. Following successful repair, he/she may return to IA.

4. Regarding transfer from pediatric cardiology to adult care

According to the comments in the guideline, this continues to be deficient and should be reinforced in pediatric cardiology with educational programs beginning at the onset of adolescence or before, as a continuous process aimed at preparing the adolescent to manage his/her own health problems as an adult. The barriers continue to be: lack of guided transfer, insufficient availability of ACHD programs, inadequate insurance coverage, deficient education of patients and caregivers regarding ACHD, inadequate resources for patients with cognitive or psychosocial impairment, lack of comprehensive case management, and different needs for evaluation and management compared to adults with acquired cardiovascular disease.

It is clear that patients live longer when they are managed by ACHD specialists. Recognition of emergencies and their appropriate care is vitally important [12]. When the guidelines are adhered to, there are fewer management errors [13]. Patients with ACHD AP classifications IB-D, IIA-D, and IIIA-D should be managed in collaboration with an ACHD cardiologist.

5. ACHD Certification Programs

In 2012, the American Board of Medical Specialties approved ACHD as a subspecialty of internal medicine ("adult") cardiology and pediatric cardiology. Therefore, for cardiologists, one marker of ACHD expertise is board eligibility/board certification in ACHD [14,15]. There are expert ACHD clinicians who are not board-certified, including those whose expertise was acquired before the development of formal certification programs and those trained outside the United States who may also have different pathways to achieve ACHD expertise. Expertise in the surgical management of patients with ACHD may be identified through board eligibility/board certification in congenital heart surgery. There are expert ACHD surgeons who are not board-certified, including those surgeons trained in other countries who are not eligible for certification in the United States. However, in the rest of America, Europe, Asia and Oceania there are programs carried out in adult congenital heart disease units within high level centers, with directed training for pediatric and adult cardiologists. Certification is required to work in the United States, but not in the rest of the countries in America. With the growing demand for specialists, it will be necessary to evaluate preparation and somehow regulate formal training in this new specialty.

6. Regarding Diagnostic Methods

As CHD complexity increases, imaging techniques become unpredictable for carrying out a correct and precise diagnosis and complete functional assessment [16-18]. Compared to the 2008 guidelines, the current recommendations add the need for intraoperative transesophageal echocardiography (TEE). From my perspective, I would also add the usefulness of TEE as a support in CHD interventionism. It is essential for guiding the procedures needed to resolve CHD residuals and sequelae [4]. The recommendations concerning avoiding the excessive use of radiation in diagnostic and therapeutic procedures are also highlighted in this guideline. In this regard, in 2015, a scientific statement on congenital heart disease in the older adult had already described the risk of various types of cancer found in adults who received cumulative radiation in childhood for various reasons.

7. Other Considerations

There is no change in the recommendations regarding exercise or pregnancy prevention; contraceptive treatment continues to have the same recommendations as the 2008 guidelines. There are contributions regarding neurodevelopment, with a mental evaluation recommended for patients (class IIa). Neurodevelopmental or neuropsychological testing may be considered in some patients with ACHD to guide therapies that enhance academic, behavioral, psychosocial, and adaptive functioning (class IIb). Mental health and neurological development problems are common in ACHD patients and may affect their quality of life. Neurological development disorders are seen more frequently in children with complex diseases. Structured professional psychological assessment may identify up to 50% more patients with mood disorders.

Depression affects an average of 42% of ACHDs, with a wide variation between continents (Asia 9 - 42%, North America 13 - 33%, Europe 9- 69%) [19]. As a result, mental disorders substantially affect the quality of life. A study carried out in Germany with 150 ACHDs showed that the prevalence of psychiatric disorders was significantly higher in ACHDs than in the general population (48.0% vs. 35.7%). Mood (30.7% vs. 10.7%) and anxiety disorders (28.0% vs. 16.8%) were the leading causes of psychiatric illness, and up to 10.7% were already receiving specific treatment for psychiatric disorders before entering the study [20].

8. Pharmacological Therapy

Progress over the last few years has continued to be deficient; the currently available medications for treating arrhythmias and heart failure have been studied and tested in the non-CHD population. The American and European clinical practice guidelines have levels of recommendation and guidelines for this type of population. So far, it has been very difficult to carry out randomized clinical trials on ACHDs. Some of the extrapolated medical treatments may end up being detrimental (III- (harm) strong, risk > benefit). It must be highlighted that there have been great advances in the treatment of pulmonary arterial hypertension associated with congenital heart disease, with the advent of endothelin receptor antagonists (bosentan, ambrisentan, macitentan) and prostanoids (treprostinil, epoprostenol, selexipag). These have been tested in several controlled, randomized studies with favorable results for ACHD.

Conclusions

In summary, the most outstanding feature of this update is the inclusion of an AP classification which will allow the severity of CHD to be classified more objectively, permitting the development of therapeutic guidelines aimed at improving the AP functional class. This had not been developed previously. Without a doubt, as the ACHD population grows, the behavior of residual lesions, sequelae and complications is discovered. The innovative techniques which have been developed today, such as hybrid procedures to palliate complex heart diseases (hypoplastic left ventricle, pulmonary atresia), will bring new knowledge regarding their behavior in adulthood. Some of these consequences are already known (liver dysfunction and cirrhosis in single ventricle physiology), but are still too limited to reach stronger conclusions.

Understanding the heterogeneity of CHDs shows us the path that not all ACHDs behave the same. The care of ACHD must be lifelong, and the number of ACHD specialists and transfer units must increase, in order to not have losses to follow up in this population.

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Conflicts of Interests

I have no conflicts of interest to declare.

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