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Congenital heart disease in women

Cardiopatías congénitas en la mujer

Ivan Romero-Rivera,* Lucelli Yáñez-Gutiérrez,‡ Yolimar Meza-Méndez,§ Igor Morr-García¶

INTRODUCTION

Advances in diagnosis, anesthesia, intensive care, and surgery have improved the evolution of congenital heart disease (CHD). Sixty years ago, only 25% survived beyond the first year of life. Currently, more than 95% survived to adulthood. It is estimated that 1/150 adults in the United States of America (USA) have different types of CHD, from subclinical forms of the bicuspid aortic valve to severe conditions; there are approximately 10 million adults with CHD in the world.

Khairy et al. analyze mortality in 71,686 patients. They observed that in 2004-2005 mortality was similar to that of the general population, quite different from the observed in 1987-1988, where high mortality was observed in the first year of life.

The estimated number of adults with CHD in the USA was 1,444,500 in 2016, a 63% increase in the estimated population with CHD since 2000. The prevalence in women was 6/1,000 and in men 3.8/1,000.

According to DiNardo, adults with CHD may: 1) have simple or complex lesions that allow survival and are seen for the first time; 2) have previous palliative procedures waiting for a new procedure; 3) be awaiting early reoperation; 4) be awaiting surgery for residual lesions; 5) be awaiting transplant; 6) also acquired heart disease. These heart diseases can be classified anatomically as type I) simple (≈ 45%), II) of moderate complexity (≈ 40%), and III) of great complexity (≈ 15%) *(Figure 1)*.

To facilitate the approach, the AHA/ACC created a functional classification: A, B, C, and D, similar to that of the NYHA for heart failure (HF), but with characteristics typical of adult CHD, indicating that patients in the classifications I B-D; II A-D and III A-D must be explored by a specialist with experience in adult congenital heart disease.

PREGNANCY

Data from the multicenter ROPAC Registry, with 57.4% of pregnant women with CHD, showed maternal mortality of 0.6% in the total group and 0.2% in those with CHD, much higher than expected (0.007% in average pregnant women).

Pre-pregnancy data, such as NYHA HF or CF signs > II, left ventricular ejection fraction < 40%, modified World Health Organization (WHO) class IV, and use of anticoagulation, were cited as predictors of HF or mortality during pregnancy.

In a review of 48 studies performed between 1983 and 2006, including 2,491 pregnant women with CHD, 34.2% have obstetric complications (2.2% embolic events, 8.7% hypertensive disease, 11% early delivery, and 3.9% premature rupture of the membranes), 11.7% of maternal cardiac complications (4.5% of arrhythmia, 4.8% of heart failure and 1.9% of other pathologies such as heart attack, stroke, etc.) and 31.4% of fetal complications among which the most important was preterm birth. CHD recurrence in children was

* Federal University of Alagoas; Santa Casa de Misericórdia Hospital in Maceió, Brazil.
‡ Cardiology Hospital. XXI Century National Medical Center, Mexico.
§ Maracay Polyclinic, Aragua, Venezuela.
¶ Professor of Medicine at the Faculty of Health Sciences of the University of Carabobo and the Central University of Venezuela, Venezuela.

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3.5%, and infective endocarditis (IE) was a rare event (0.5%).

The number of pregnancies in patients with CHD increases rapidly. Data from the US National Inpatient Sample from 1998-2007 show that the number of deliveries in patients with CHD increased by 34.9%, compared to an increase of 21.3% in the general population.

The care of a pregnant woman with CHD involves:

1. Preconception: women should be assessed before planning a pregnancy using risk stratification scales (ZAHARA, CARPREG, and WHO). The modified WHO risk stratification should be used initially *(Table 1)*.

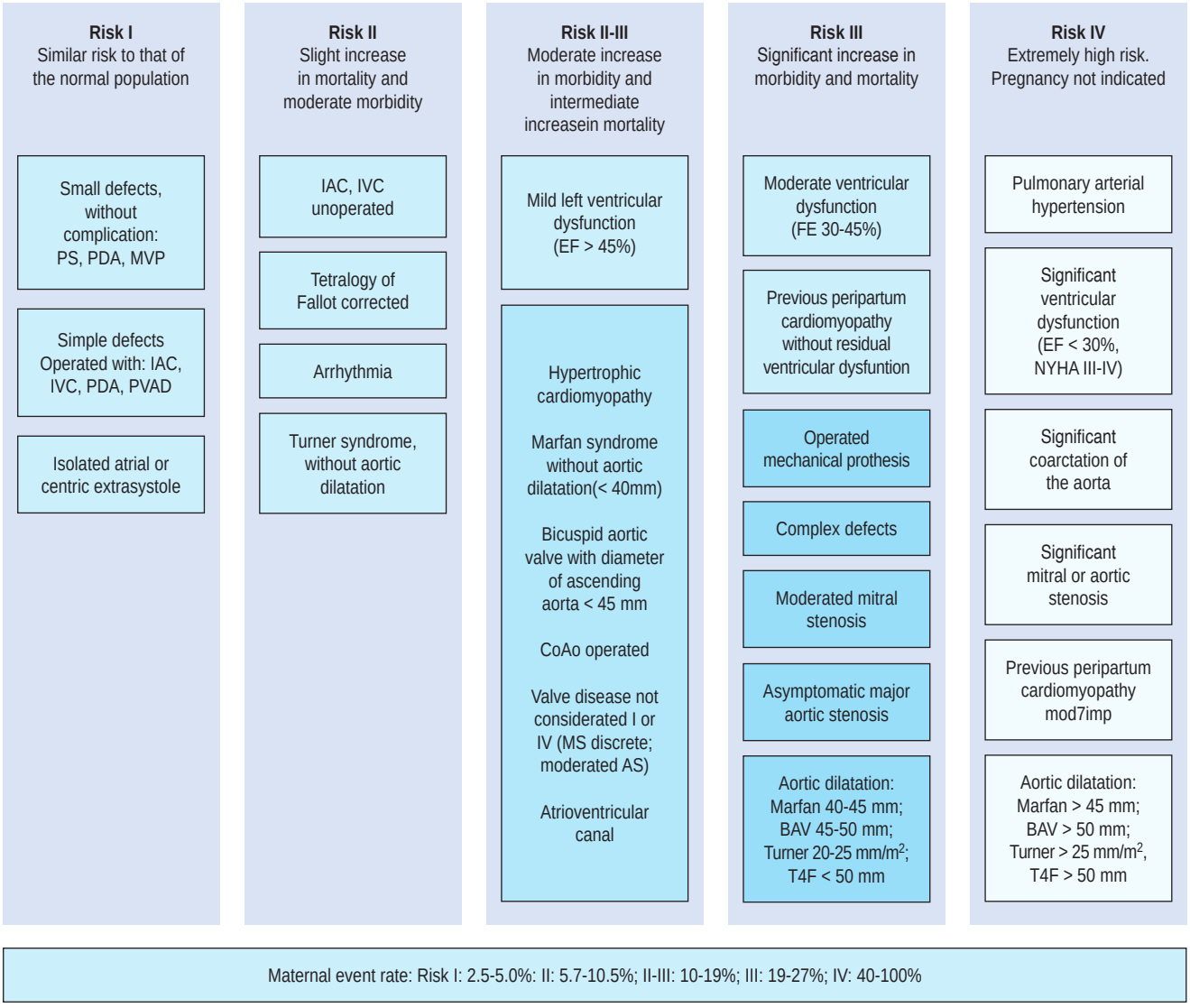

Figure 1: Anatomical classification of congenital heart diseases in adults.
PS = pulmonary stenosis. PDA = patent ductus arteriosus. MVP = mitral valve prolapse. IAC = interauricular communication. IVC = interventricular communication. PVAD = percutaneous ventricular assist devices. CoAo = coarctation of the aorta. MS = mitral stenosis. AS = aortic stenosis. BAV = bicuspid aortic valve; T4F = Tetralogy of Fallot. EF = ejection fraction. NYHA = New York Heart Association Classification.
Modified from: Warnes CA et al.[1], Warnes CA et al.[2], Khairy P et al.[3]

Table 1: Cardiovascular risk according to the WHO in women with congenital heart disease, follow-up strategy, and estimated mortality.

I. Simple congenital heart disease
- Congenital aortic or mitral valve injury isolated (except MV parachute, cleft)
- Isolated small IAC e IVC
- Pulmonar stenosis, small
- Simple defects operated: IAC, IVC, PDA, no residual injury or sequel

II. Congenital heart disease of moderate complexity
- Aortoventricular tunnel
- Abnormal drainage of the pulmonary veins
- AOCA of pulmonary artery or Ao (opposite sinus)
- Atrioventricular septal defects (partial or complete)
- Congenital mitral or aortic stenosis
- Aortic coarctation
- Ebstein anomaly
- Right ventricular outflow tract obstruction
- IAC ostium primum or sinus venosus
- Moderate or significant IAC unoperated
- Moderate or significant ductus arteriosus
- Moderate to severe pulmonary reflux
- Moderate or severe pulmonary stenosis
- Stenosis of peripheral branches of the pulmonary artery
- Sinus of Valsalva aneurysm/fistula
- Sub or supra valvular aortic stenosis (except HMO)
- Atrioventricular valve in straddling corrected Tetralogy of Fallot
- IVC with associated defects and/or mod/imp shunt
- Right ventricle with double chamber
- Marfan syndrome (inheritance of alt Ao), Turner
- Sub, valvar and supra mod/imp pulmonary stenosis
- TGA after switch surgery

III. Congenital heart disease of great complexity
- Cyanotic congenital heart disease (unoperated or palliated)
- Double exit pathway of the ventricles
- Fontan surgery
- Interruption of the aortic arch
- Mitral atresia
- Single ventricle
- Pulmonary Atresia (all forms)
- Transposition of the great arteries (TGA). TGA except patients with previous switch surgery
- Truncus arteriosus
- Others: criss-cross heart; isomerism, heterotaxic syndromes, ventricular inversion
- Congenital heart disease associated with pulmonary hypertension, including Eisenmenger syndrome

MV = mitral valve. IAC = interauricular communication. IVC = interventricular communication. PDA = patent ductus arteriosus. AOCA = anomalous origin of coronary artery. TGA = transposition of great arteries. Ao = aorta.
It is modified from: Drenthen W et al.[8]

2. During pregnancy: Multidisciplinary care to diagnose and treat possible complications. Guide the use of cardiovascular drugs with effects on the fetus.
3. In childbirth: except for some heart diseases, the indication for delivery is obstetric. It is essential to prevent complications related to pain, anxiety, and massive venous return when decompressing the pregnant uterus or using thromboprophylaxis.
4. Puerperium: divided into immediate (24 hours), mediate (7 days), and late (42 days), will be followed up to one year after delivery or interruption of pregnancy. Guiding family planning without interfering with the use of cardiovascular drugs. The physiological changes of pregnancy tend to normalize 45 days after delivery, requiring cardiovascular evaluation at this time.

COMPLICATIONS

Patients with CHD generally use medication for life and require multiple surgeries and hospital admissions; they can present a higher number of complications or mortality during pregnancy, severe arrhythmias and heart failure, and reduced life expectancy. We cannot forget acquired diseases, such as systemic arterial hypertension, acute myocardial infarction, and cerebral vascular accident.

IE is more frequent than in the general population and more severe in prosthetic valves, including those with percutaneous implantation, valve repair with a prosthetic ring, previous IE, any cyanotic CHD or repaired with prosthetic material, up to 6 months after the procedure or permanent if a residual shunt or valvular insufficiency persists.

Antibiotic prophylaxis is recommended in patients at high risk for IE, even during childbirth, without consensus on the preventive use of antibiotics in all women with CHD.

Bleeding diathesis and thrombosis are frequent complications, and their treatment and prophylaxis contribute to survival. Spontaneous bleeding is mild and self-limited (dental, epistaxis, bruising, menorrhagia). Hemoptysis is the external manifestation of intrapulmonary hemorrhage, common in Eisenmenger syndrome.

Concerning thrombosis, the associated risk factors are female gender, oxygen desaturation, senility, biventricular dysfunction, dilatation of pulmonary arteries, and post-surgical complications such as Fontan, in which there may be total or partial thrombotic obstruction of the shunt cavopulmonary. Its treatment includes thrombectomy, percutaneous angioplasty (stent), surgery, and anticoagulant therapy or thrombolysis.

Strokes are frequent and related to thromboembolic events, microcytosis, endothelial dysfunction, and paradoxical emboli due to endocavitary electrodes and catheters. Anticoagulation is not routinely indicated in patients with Pulmonary Hypertension, and its prescription is individualized, as in the case of mechanical valves, vascular prostheses, supraventricular arrhythmias, presence of thrombosis or pulmonary embolism, provided that the bleeding risk is low.

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Correspondence:
Ivan Romero-Rivera
E-mail: irrivera@uol.com.br