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Editorial

Adult congenital heart disease in Spain: present situation and future perspectives

Cardiopatías congénitas del adulto en España: situación actual y perspectivas futuras

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Due to the development of various congenital heart disease (CHD)-related disciplines in recent decades, particularly, surgery in early infancy, more than 90% of newborns with these conditions reach adulthood.[1] The subsequent dramatic increase in the adult population with CHD treated in infancy means that this population already exceeds that of infants with CHD in absolute numbers.[2] Because CHD is present in about 1 in every 100 live births, an estimated 2.3 million European adults have such a condition.[3]

The diagnosis, management, and follow-up of patients with adult CHD (ACHD) clearly have specific characteristics, often straddling those of infant and adult cardiology. Accordingly, the numerous guidelines and recommendations on the management of ACHDs published in recent decades in Europe and North America[4–6] agree that these patients should be managed in experienced centers with appropriately trained staff. In particular, specialized care is most beneficial for patients with moderate-to-high complexity CHD. Appropriately organized care of patients with ACHD in specialized centers and by specifically trained professionals has a huge impact on survival,[7] which is why integrated management of this population and specialization of the medical staff attending these patients are essential to meet their health care needs.

Canada was the first country to create a national network of specialized centers for the care of patients with ACHD. Many other countries with a tradition of treating ACHDs have been inspired by this model, which is based on the creation of multidisciplinary teams dedicated to the integrated care of ACHDs. These teams are located in centers organized in different levels, coordinate among themselves, and comprise professionals with specific training. This model has been implemented to greater or lesser extent by several European countries, such as the United Kingdom, the Netherlands, Switzerland, and Germany, according to their demographic and socioeconomic characteristics and health care systems. In Germany in particular, the creation of a national care network for ACHDs was driven by national scientific societies for adult cardiology, pediatric cardiology, and cardiac surgery, who endeavored to organize a care network of accredited centers for ACHD management.[8] In addition, considerable emphasis has been placed on the accreditation of the cardiologists treating this population and ACHDs have been established as a new subspecialty of both pediatric and adult cardiology.[9]

However, the development of the subspecialty in European countries as a whole has not followed a steady rhythm. In 2010, Moons et al.[3] published the results of a survey of the state of ACHD care in Europe that revealed considerable differences among countries. In 2014, in an attempt to standardize the health care organization for this population, the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology proposed minimum criteria for the material and human resources that should be possessed by centers or units dedicated to the treatment of ACHDs, as well as the training required for the ACHD subspeciality.[10]

In this context, Spain has undergone the same epidemiological changes in CHDs. Although no population-based data are yet available, extrapolation from the results of various prevalence studies suggests that about 120 000 adults have a CHD.[11] In response to this population increase, recent years have seen the creation of ACHD centers. These units have been added to the few large centers with a recognized tradition of treating these patients, largely the result of the individual initiatives of certain professionals with a special interest in the field.

From this point in the development of the subspecialty, we need to progress toward an appropriate planning of resources—not just intuitively, but also in line with international recommendations—to optimize the care of ACHDs in Spain as much as possible. However, no objective data were previously available on the structure of ACHD management in Spain or on how many and what type of patients are undergoing follow-up in specialized centers.

In a recent article published in *Revista Española de Cardiología*, Oliver-Ruiz et al.[12] present a comprehensive analysis of the organizational structure and health care activity of centers treating patients with ACHD in Spain, as well as the clinical characteristics of the patients treated in these centers. In 2014, the authors administered a survey to Spanish centers treating ACHD patients, selected those with at least 1 specialized clinic, and divided them into 2 groups (levels 1 and 2) based on their health care structure. In total, 24 centers had at least 1 specific ACHD clinic; 10 were considered to be level 1 because they had a structure able to meet a*l*l of the needs of patients with ACHD. International consensuses[13] estimate that a level 1 national center is required to manage a population of 3 to 10 million people. It is also estimated under this model that there should additionally be a regional or level 2 center for every 2 million inhabitants; this unit would refer patients with specific needs to level 1 centers in the network. The

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data from Oliver-Ruiz et al.[12] are hugely valuable because, as noted by the authors, they indicate 1 specialized center for every 2 million people and 1 level 1 center for every 4.7 million, which is in line with the current recommendations.[10] Notably, the vast majority of level 2 centers began their ACHD activity in the last 10 years, with a median duration of activity of 9 years. This indicates that Spain is increasingly recognizing the value of specific care for patients with ACHDs, so much so that the Spanish density of specialized centers has rapidly reached that of other European countries, particularly due to the creation of level 2 centers. However, and in contrast to other countries, the study data reveal an uneven geographical distribution of these centers, with a pronounced concentration of level 1 centers in large cities and, above all, extensive regions without specialized care centers. Given that the survey used as a basis for the study was performed in 2014, the data might be incomplete, as mentioned by the authors, with recently established specialized centers omitted from the study. Regardless, their findings prove that improvements are possible in the distribution of centers able to provide specialized care to ACHD patients in Spain. Moreover, the results are invaluable for guiding a fairer distribution of health care resources and structure throughout Spain.

Notably, although the level 1 centers had more patients under follow-up than the level 2 centers, the total number of patients under follow-up in the centers analyzed by Oliver-Ruiz et al.[12] was about 20 000. The above-mentioned estimate of the total number of ACHD patients in Spain (about 120 000) indicates that the overwhelming majority of Spanish patients with ACHD are not being followed up in specialized centers. This problem is not unique to Spain: some population data-based studies have estimated that only 30% of ACHD patients are under follow-up in specialized centers such as those recommended in international guidelines, with similar percentages in countries with universal health care systems similar to that of Spain, such as Canada,[14] and most European countries.[15] Although the reasons for the huge number of patients not under follow-up in specialized centers are multifactorial, the uneven national distribution of these centers has undoubtedly been a contributing factor.

Another important result provided by Oliver-Ruiz et al.[12] is the evidence that the structure, facilities, and staff of Spanish centers are in line with those of international recommendations.[10] However, there is a deficit in the incorporation of specialized nursing staff and in structured transition programs from pediatric cardiology departments.

The recommendations[10] include the incorporation of nursing staff into ACHD units. However, due to the heterogeneity of the specific training programs for nurses in Europe and partly because the funding of nursing resources can sometimes be more difficult to justify economically, the incorporation of these professionals into ACHD programs in Spain has not been made a priority. Again, this problem is not unique to Spain. As shown in a recent study analyzing the characteristics of 96 European ACHD clinics,[15] there is huge variability among centers in both the number of patients under follow-up and in human resources. Strikingly, just 4% of centers fully met the staffing recommendations proposed in the above-mentioned consensus document from the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology,[10] largely because most clinics lack the nonphysician health care workers recommended in this document (psychologists, social workers, or specialized nurses).

However, while this same study revealed that 89% of European centers do have a specific transition program,[15] the study by Oliver-Ruiz et al.[12] showed that only 30% of Spanish centers have such a program. As noted by the authors, and in contrast to other European countries, most Spanish health care transition programs for CHD have traditionally been handled by nursing staff. Evidence suggests that the transition period from pediatric to adult health care can account for up to 50% of losses to follow-up among patients with CHD, particularly among those with stable disease when they reach adulthood.[14] To avoid the potential consequences of these follow-up interruptions, it is essential to promote transition programs in ACHD units.

To analyze the clinical characteristics of patients under follow-up in centers specialized in the care of ACHD, the authors used data from a cross-sectional registry of clinical activity from 18 of the 24 centers (level 1, 7; level 2, 11) over 2 months in 2017. In total, 32% of the included patients had simple heart disease, 44% had moderate disease, and 24% had highly complex disease. The results reveal that the vast majority of interventional procedures in patients with ACHD are performed in level 1 centers and that these centers predominantly undertake the follow-up of moderate and highly complex patients. However, the study shows that 17% of patients under follow-up in level 2 centers have highly complex CHD and that about a quarter of therapeutic procedures undertaken in patients with ACHD are performed in these centers. This classification of heart diseases as simple, moderate, and complex is controversial because some CHD patients classified as simple would benefit from an evaluation in centers with experience with particular situations, such as the coexistence of ostium secundum atrial septal defect and pulmonary hypertension. Generally, patients with highly complex CHD benefit from follow-up, at least shared, in more experienced centers. The results of the present study largely reflect this situation, in agreement with the current recommendations.[10] Under this model, patients with ACHD should be evaluated at least once in a level 1 center to determine the frequency and health care level of the follow-up. However, as indicated by the authors, therapeutic procedures should be concentrated in the centers with most experience. While this recommendation is clear for more complex heart diseases, it is also important for many of the heart diseases considered simple, in which determination of the appropriate procedure is sometimes more complex than the procedure itself. In this context, the development of functional collaborative networks among centers of different health care levels is crucial to provide adequate care to patients with ACHDs.

The work by Oliver-Ruiz et al.[12] is of tremendous interest because it gives us an overall view of the current health care situation for patients with ACHD in Spain and provides hugely valuable data on the organization of the health care structure, clinical activity, and characteristics of patients under follow-up in specialized ACHD units in Spain. The study confirms the considerable advances in the speciality in Spain, as well as the aspects requiring specific attention. The information obtained will be vital for resource planning aimed at improving the collective outcomes of patients with ACHD, a rapidly growing population.

CONFLICTS OF INTEREST

None declared.

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