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Congenital Heart Disease and Pregnancy Priorities:

Balancing Risks and Hopes*

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Women rarely make decisions about pregnancy based on its physical and physiologic consequences alone. These opinions evolve over a woman's lifetime and are influenced by society, her peers and partners, and stories she hears from other women and, in this era, from social media. Women with congenital heart disease (CHD) are no different in this regard. As medical and surgical advances have improved outcomes in our CHD patients, many perceive pregnancy as just another normal milestone that they can accomplish.

Historically, pregnancy counselling for adult CHD (ACHD) women has not been consistent. As ACHD physicians, we have anecdotes of women who have undergone reproductive sterilization for simple cardiac defects. Many ACHD women have stories of successfully carrying multiple pregnancies after defying their physicians' recommendations. These experiences have caused a mistrust in the "expertise" that our medical opinions offer.

Over the years, systematic studies have been adapted into risk scores[1–3] and guidelines[4] to help physicians and their ACHD patients make decisions about pregnancy. Yet, even within these guidelines, there are significant knowledge gaps such as those pertaining to mixed lesions and long-term outcomes of pregnancy with CHD. Numerous studies have shown that the frequently used pregnancy risks scores such as CARPREG (Cardiac Disease in Pregnancy) and ZAHARA (Zwangerschap bij Aangeboren HARtAfwijking) have limitations at predicting outcomes.[5–9] Additionally, the true risk of pregnancy in a woman with CHD

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is not defined by the severity of her cardiac lesion alone but the expertise offered by her care center in managing the complications that arise. While most centers have cardiac anesthesiologists, interventionalists, surgeons, obstetrician, maternal fetal medicine experts, and neonatologists, their experience in working together as a team and providing the needed interventions in a timely manner largely defines outcomes in these patients.

Hence, every ACHD physician who cares for pregnant women will agree that while all uncomplicated pregnancies are alike, every high-risk ACHD pregnancy is complicated in its own way.

ACHD women and their physicians are most concerned about maternal death during pregnancy, which in most cases can be avoided with unrestrained use of health care resources. Instead, women develop hypertension, arrhythmias, heart failure, and thromboembolic complications[8] which, while treatable, can change their risk of long-term cardiovascular comorbidities and quality of life. Furthermore, exacerbation of CHD sequalae can lead to neonatal consequences such as premature birth, developmental abnormalities, intrauterine growth retardation, as well as neonatal death.[8,10] Similar to pregnancy, the burden of these outcomes is perceived differently by different women.

Hence, pregnancy counselling in a high-risk ACHD woman should be tailored to the patient, in the context of available clinical resources and with the understanding that her beliefs and concerns may be very different than what we assume as physicians.

In the paper published in this issue of *JACC: Advances*, Herrick et al[11] provide a roadmap that should guide our conversations while counselling ACHD women about pregnancy. The authors identified 5 unique subgroups of ACHD women based on their beliefs and priorities. While there is some overlap in belief systems between groups, the study shows that ideologies related to reproductive decisions span a wide spectrum, ranging from women who feel strongly about getting pregnant irrespective of the risk their CHD imparts (group 1) to women who do not want to impose undue risk to their health and will not consider pregnancy unless all modifiable risk factors are addressed (group 5). While some women are strongly guided by their personal values related to contraception and pregnancy termination (group 4), most women struggle with anxiety related to limited understanding of their CHD. This anxiety is primarily driven by how their CHD affects the short-term outcomes (group 2) and long-term outcomes for themselves and their children (group 3).

While their personal beliefs may be set, by participating in the study, these women reveal that they are looking to partner with their ACHD physicians to achieve the best possible outcomes. Hence, counselling ACHD women regarding pregnancy should be a conversation about how their beliefs and priorities shape the acceptable risk that the patient is willing to undertake for herself and her baby, rather than an ultimatum that challenges their well-established personal beliefs and priorities.

At this time, it is unknown if neonatal outcomes such as prematurity, intrauterine growth retardation, developmental abnormalities, or death influence pregnancy decisions among ACHD women and are not included in the study. However, it is an important part of pregnancy counselling as these outcomes occur frequently in high-risk pregnancies.[8,10]

Additionally, the study does not investigate how patients in each of the subgroups respond to different counselling techniques. However, the authors do provide suggestions on how counselling can be tailored to each of these patients. While most patients would benefit from conversations related to cardiac optimization prior to pregnancy, the authors point out that patients who have significant anxiety related to their CHD may also benefit from psychological counselling.

Another novelty related to the study is the use of Q-methodology to collect and analyze women's perceptions related to pregnancy. Q-methodology is a mixed methodology approach that subjectively collected patients' viewpoints on pregnancy by ranking statements related to their reproductive ideology.[12] Following this survey, their cumulative responses are grouped based on similarities in ideology, thereby establishing the 5 distinct subgroups described in their manuscript. This approach is being increasingly used in health care research but is novel within the ACHD field. Since decisions related to pregnancy can be emotionally charged, the use of a structured multiquestion survey to rank reproductive priorities provided a neutral yet powerful avenue for understanding various viewpoints of women with CHD. Of the 178 women surveyed, there was a wide variation in numbers in each group ranging from 12 to 55. Interestingly, 22 participants did not identify with any single group, which invites the possibility that other, yet unknown, priorities may be driving decisions related to pregnancy. Alternatively, since wording or the sequence in which questions are asked can influence responses, further curating the questions in subsequent studies may provide a more comprehensive understanding of this topic.

As physicians, while we strive to deconstruct the clinical implications associated with CHD and translate it to risk for our patients, the study shows that in a substantial proportion of cases, women have already formed their opinions about pregnancy in the context of their heart disease. Hence, as health care providers, our efforts should focus on understanding these views related to pregnancy, identifying modifiable risk factors, defining acceptable risks, and engaging multidisciplinary physicians including mental health providers who can intervene early to prevent adverse outcomes, consequently building trust between women with ACHD and their medical team.

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REFERENCES

1. Silversides CK, Grewal J, Mason J, et al. Pregnancy outcomes in women with heart disease: the CARPREG II study. J Am Coll Cardiol. 2018;71:2419–2430. [PubMed: 29793631]
2. Siu SC, Sermer M, Colman JM, et al. Prospective multicenter study of pregnancy outcomes in women with heart disease. Circulation. 2001;104:515–521. [PubMed: 11479246]
3. Drenthen W, Boersma E, Balci A, et al. Predictors of pregnancy complications in women with congenital heart disease. Eur Heart J. 2010;31:2124–2132. [PubMed: 20584777]
4. Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, et al. 2018 ESC guidelines for the management of cardiovascular diseases during pregnancy. Kardiol Pol. 2019;77:245–326. [PubMed: 30912108]

5. Pijuan-Domenech A, Galian L, Goya M, et al. Cardiac complications during pregnancy are better predicted with the modified WHO risk score. Int J Cardiol. 2015;195:149–154. [PubMed: 26043149]

6. Denayer N, Troost E, Santens B, et al. Comparison of risk stratification models for pregnancy in congenital heart disease. Int J Cardiol. 2021;323:54–60. [PubMed: 32931856]

7. Kim YY, Goldberg LA, Awh K, et al. Accuracy of risk prediction scores in pregnant women with congenital heart disease. Congenit Heart Dis. 2019;14:470–478. [PubMed: 30729681]

8. Khairy P, Ouyang DW, Fernandes SM, Lee-Parritz A, Economy KE, Landzberg MJ. Pregnancy outcomes in women with congenital heart disease. Circulation. 2006;113:517–524. [PubMed: 16449731]

9. Balci A, Sollie-Szarynska KM, van der Bijl AG, et al. Prospective validation and assessment of cardiovascular and offspring risk models for pregnant women with congenital heart disease. Heart. 2014;100:1373–1381. [PubMed: 25034822]

10. Hardee I, Wright L, McCracken C, Lawson E, Oster ME. Maternal and neonatal outcomes of pregnancies in women with congenital heart disease: a meta-analysis. J Am Heart Assoc. 2021;10:e017834. [PubMed: 33821681]

11. Herrick N, Al-Rousan T, Rodriguez C, et al. Priorities and understanding of pregnancy among women with congenital heart disease: a mixed-methods study. JACC Adv. 2022;1(4):100112.

12. Churruca K, Ludlow K, Wu W, et al. A scoping review of Q-methodology in healthcare research. BMC Med Res Methodol. 2021;21:125. [PubMed: 34154566]