Cardiology in the Young (2017), **27**, 236–242
doi:10.1017/S104795111600038X

Original Article

Current attitudes and clinical practice towards the care of pregnant women with underlying CHD: a paediatric cardiology perspective

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Abstract *Objectives:* The growing number of women with CHD presents unique challenges, including those related to pregnancy, which can lead to significant morbidity and mortality. We sought to evaluate the perception of paediatric cardiologists towards the reproductive health of women with CHD. *Methods:* Paediatric cardiologists in the United States of America were invited to participate in a cross-sectional, anonymous survey. Information solicited included knowledge of contraceptive methods, experience caring for pregnant women with CHD, and referral patterns including the utilisation of high-risk obstetric and adult CHD specialists. *Results:* A total of 110 cardiologists responded – 90% with an academic affiliation and 70% with ⩾10 years' clinical experience. Although 95% reported an understanding of available contraceptive options, 32% did not feel comfortable recommending birth control. Pregnant women with CHD were seen by 83% of responders, and 37% of the responders reported a low level of comfort in doing so. Among all respondents, 73% indicated that they would refer a pregnant CHD patient to a high-risk obstetrician and 60% to an adult CHD specialist – almost all respondents would not transfer care to a non-adult CHD cardiologist. Among paediatric cardiologists, 81% indicated that they would resume their patient's care following delivery. *Conclusion:* Our results illustrate a gap in what physicians feel should be done and the care that they feel comfortable providing pregnant women with CHD. As this population continues to grow, training adult CHD cardiologists with specific skills in reproductive health in women with CHD is the first step to closing the care gap that exists in the management of such patients.

Keywords: CHD; adult(s) with CHD; pregnancy

Received: 25 October 2015; Accepted: 5 March 2016; First published online: 11 April 2016

The incidence of CHD worldwide is slightly >1% of all live births.[1] With extensive medical and surgical innovation over the past 50 years as well as high-quality care received in childhood, 85–90% of children with CHD in the United States of America can be expected to survive into adulthood.[2–4] As a result, the number of adults with CHD in the United States of America has reached an estimated 1.5 million with increasing numbers every year.[2–5] Studies have also estimated that the proportion of pregnant women with underlying CHD also continues to expand.[3]

The growing number of adult CHD patients has presented unique challenges for practitioners ranging from access to healthcare, non-cardiac and cardiac surgery, ongoing psychosocial care needs, and obstetrical care. Risk factors for complications during pregnancy in women with CHD include cyanosis, management of anticoagulation in the setting of mechanical valves, paradoxical thromboembolic events, aortic dilation, pulmonary hypertension, both systemic and pulmonary ventricular dysfunction, and exacerbation of arrhythmia.[6] The importance of these

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hazards has led to the establishment of multiple guidelines to help provide a framework for risk assessment for pregnancy as well as contraceptive recommendations in both general and lesion-specific terms.[7–11] Challenges that remain include identifying the appropriate cardiology physician who is adequately trained to care for pregnant women with mild-, moderate-, or great-complexity CHD as well as identifying the ideal location where management of issues related to pregnancy, delivery, and any necessary procedures can be performed.

To date, there have been limited evaluations of healthcare delivery to pregnant women with CHD. Although more than 110 institutions from across the United States of America and Canada have worked to develop formal adult CHD programmes to meet the distinct needs of this population, it still remains unclear as to where and who should be providing the care for pregnant women with CHD.

Methods

In 2011–2012, we invited 199 paediatric cardiologists from across the United States of America to participate in a cross-sectional, anonymous survey. The survey was delivered via e-mail, and was re-sent up to a maximum of three times to non-respondents over a 6-month period. The survey was sent via e-mail to paediatric cardiologists from all parts of the United States of America and some parts of Canada via a central online list server. The survey consisted of items regarding basic programme demographics such as size, number of surgeries per year, and setting as well as the programme's current model for the care of women with CHD (Appendix). Included were questions regarding knowledge of contraceptive methods, experience caring for pregnant women with CHD, and referral patterns including the utilisation of high-risk obstetric and adult CHD specialists. Each participant's responses remained anonymous and were transmitted directly into an online, secure database.[12]

Results

A total of 111 cardiologists responded to the survey. Nearly all (90%) were affiliated with an academic affiliation and 70% had ⩾10 years' experience. More than 95% had previously reported the presence of an adult CHD clinic at their institution as part of the 2011 Adult Congenital Heart Association clinic directory.[13] Although 95% reported an understanding of available contraceptive options, 32% did not feel comfortable recommending birth control to women with CHD. Nearly all respondents (98%) discussed pregnancy with their patients and most often (32%) when they were 16 years of age. Most (96%) indicated that they would counsel women with the following medical conditions to avoid pregnancy: pulmonary hypertension (93%), aortic aneurysm with underlying connective tissue disease (77%), and Fontan palliation (71%).

Among all, 83% of the respondents encountered pregnant women with CHD in their practice. Upon learning that their patient was pregnant, 60% indicated they would refer her to see an adult CHD specialist and 73% would also refer her to a high-risk obstetrician. Nearly all respondents (98%) indicated they would not transfer the care of their pregnant patient to an adult cardiologist without dedicated adult CHD training. Although 71% of the respondents had followed-up a woman throughout her entire pregnancy, with 95% within the past 5 years, 35% reported a low level of comfort in doing so. Notably, 81% of respondents stated that they would resume their patient's primary cardiovascular care following delivery and not transition to an adult CHD specialist.

Discussion

Our results illustrate a gap in between what physicians feel should be done and the care that they feel comfortable providing pregnant women with CHD. This discrepancy suggests that providers view pregnancy as a complicated physiological state, but not necessarily one that heralds the need for transition of care to an adult CHD specialist. Given that 35% of those surveyed reported a low level of comfort in providing care throughout pregnancy, further research is necessary among the healthcare community not only to educate the patients but also to educate healthcare providers.

Pregnant women with CHD are increasingly being seen in paediatric cardiology and obstetrical offices and, although cardiac disease complicates a small percentage of all pregnancies in developed countries (1–4% in the United States of America), it is a major cause of non-obstetric maternal morbidity and mortality.[14] Historically, rheumatic heart disease was the most common form of cardiac disease encountered in pregnant women;[15] however, CHD has become the most common form of heart disease complicating pregnancy in the modern world.[16] Many women, including those with CHD, are also postponing childbearing until later in life. Indeed, the average age of the first live birth in the United States of America has recently risen to 26 years.[17] With advancing maternal age, other co-morbidities such as hypertension, diabetes, hypercholesterolaemia, and even coronary artery disease have become more common and have increased the

https://doi.org/10.1017/S104795111600038X Published online by Cambridge University Press

incidence of acquired heart disease complicating pregnancy.[3,12]

Pregnancy in women with CHD can lead to cardiac complications during pregnancy and their risk can be estimated by assessing a number of factors. The "CARPREG Score" is based on the single largest prospective study of 562 women with congenital or acquired cardiac disease or arrhythmias who had 617 pregnancies.[10] The actual rate of primary cardiac events such as pulmonary oedema, arrhythmia requiring treatment, stroke, cardiac arrest, or death was 13% overall, with 55% occurring antepartum. Risk factors for cardiac events during pregnancy included poor functional class (NYHA class II to IV), cyanosis, previous cardiac event such as heart failure, transient ischaemic attack, stroke, or arrhythmia, left heart obstruction (mitral valve area of $<2\,cm^2$, aortic valve area of $<1.5\,cm^2$, peak left ventricular outflow gradient >30 mmHg), and left ventricular systolic dysfunction (left ventricular ejection fraction $<40\%$).[10] The ZAHARA investigators have another model for predicting pregnancy complications in women with CHD. In addition to cyanosis, need for cardiac medications before conception, and left heart obstruction, other independent risk factors included mechanical valve replacement and systemic or pulmonary atrioventricular valve regurgitation related with the underlying complex CHD.[9] More recently, the modified World Health Organisation guidelines created a four-category schema from which risk assessment could be derived, ranging from minimal risk to pregnancy contraindicated. These guidelines promote individualised plans of care and are regarded as among the most accurate in predicting morbidity.[11]

There are many challenges facing practitioners who care for adult CHD population, such as providing appropriate antenatal and peripartum management of pregnant women with CHD, the optimal location to provide care and obtain necessary procedures, and securing the expertise of and collaboration with non-cardiac subspecialties such as high-risk obstetrics and anaesthesia to assist with the management of labour, delivery, and any complications that may arise in the peripartum or postpartum period.[18] Therefore, it must be appreciated that not all physicians or hospitals are equipped to care for pregnant women with CHD, particularly if complications arise, which may be adult-onset cardiac or non-cardiac in nature.[16,19–23]

As this study illustrates, the majority of paediatric practitioners care for women with CHD who are of childbearing age. Multiple studies have created various schemas to assess risk to the fetus and gravid mother, but none of them is perfect. In the future, larger studies hopefully can be conducted to more accurately risk stratify patients. In addition, future enquiries into clinical practice patterns might provide useful data regarding where these patients are cared for, by whom, what level of counselling they receive, and why.

Limitations

Our study has several important limitations. First, only 111 paediatric cardiology physicians responded across North America. We are also unable to provide detailed information regarding the non-responders. As a result, it is possible that the data reported in this study do not reflect the practices of all paediatric cardiologists currently in practice either in universities or private settings. Another limitation of the survey methodology is recall bias on the part of the provider. Finally, we limited our survey distribution to paediatric cardiology practitioners; therefore, our results may be falsely skewed towards patients remaining with the primary paediatric provider.

Conclusions

Even with tremendous advances in recent years, patients with CHD continue to face the prospect of both surgical and percutaneous interventions, arrhythmia, heart failure, and even premature death. This fact highlights the need of adult CHD patients to maintain lifelong cardiac care from cardiologists with appropriate training and expertise. This is particularly true during pregnancy. Best care practice for these patients requires a team approach including specialists in adult CHD cardiology, maternal–fetal medicine, and obstetrical anaesthesiology. Although the 32nd Bethesda Conference and the 2008 ACC/AHA guidelines on the care of adult CHD provide management strategies for the practicing cardiologist, we present a disparity between published guidelines and the reality of care for pregnant women with CHD. The potential implications of the findings from this survey include providing a continuation of unmet healthcare needs in pregnant women with CHD as well as insufficient attention to both adult-onset cardiac and non-cardiac diseases complicating pregnancy. Training adult CHD cardiologists with knowledge about reproductive health in women with CHD is one of the first steps to closing this care gap.

Acknowledgements

None.

Financial Support

This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Conflicts of Interest

None.

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Appendix

Baseline demographic questions for the participant:

1. **Gender:** Male [ ] Female [ ]
2. **Number of years since completing cardiology training**

   <5 years [ ] 6-10 years [ ] 11-15 years [ ] 16-20 years [ ] 21-25 years [ ] > 25 years [ ]
3. **Primary specialty**

   Pediatric Cardiology [ ]

   Adult Cardiology [ ]

   Both [ ]
4. **Cardiology sub**-specialty

   Adult Congenital Heart Disease [ ]

   Cardiac Imaging [ ]

   Cardiac Intensivist [ ]

   Heart failure / Transplant [ ]

   Interventional cardiology [ ]

   Electrophysiology [ ]

   Other (please specify): [ ]
5. Region of North America in which you practice:

   Northeast [ ]

   Mid West [ ]

   West Coast [ ]

   Southeast [ ]

   Southwest

   Canada [ ]

   Other (please specify)

Contraception Questions:

6. **Are you aware of the available birth control options for women of child-bearing age?**

   Yes [ ] No [ ]
7. **Do you feel comfortable recommending birth control to women of child-bearing age with congenital heart disease (CHD)?**

   Yes [ ] No [ ]
8. **Do you offer birth control advice to patients without him/her asking inquiring first?**

   Yes [ ] No [ ] Rarely [ ]
9. **For your FEMALE patients, do you routinely discuss the risk of transmission of CHD to their offspring?**

   Yes [ ] No [ ] Rarely [ ] Only if syndromic [ ]
10. **For your MALE patients, do you routinely discuss the risk of transmission of CHD to their offspring?**

    Yes [ ] No [ ] Rarely [ ] Only if syndromic [ ]

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Pregnancy Related Questions:

11. At what age do you typically discuss pregnancy with your female patients?

<14 years [ ] 15yr [ ] 16yr [ ] 17yr [ ] 18yr+ [ ] Never [ ]

12. Do you encounter women with underlying CHD who are pregnant in your practice?

Yes [ ] No [ ]

13. Have you followed a woman with CHD through her *entire* pregnancy?

Yes [ ] No [ ]

14. If your answer to Qs. 11 was 'Yes', when was the last time you followed a woman with CHD through her *entire* pregnancy ?

<5 years [ ] 6-10 years [ ] 11-15 years [ ] 16-20 years [ ] 21-25 years [ ] > 25 years [ ]

15. **Do you feel comfortable managing females with CHD during pregnancy?**

Yes [ ] No [ ]

16. Would you transfer a woman with CHD who is considering pregnancy to an Adult Congenital Heart Disease (ACHD) specialist, *PRIOR* to her getting pregnant?

Yes [ ] No [ ] We do not have an ACHD specialist [ ]

17. Would you transfer a woman with CHD to an ACHD specialist *AFTER* she becomes pregnant?

Yes [ ] No [ ] We do not have an ACHD specialist [ ]

18. Would you consider transitioning a patient with CHD to an adult *(solely Internal Medicine trained)* cardiologist (i.e. not an ACHD Specialist) upon learning that she is pregnant?

Yes [ ] No [ ]

19. If your response to Qs. 18 was yes, please describe the lesions that you feel would be appropriate for an adult (*solely Internal Medicine trained*) cardiologist (i.e. not an ACHD Specialist) to manage:

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20. Would you counsel a woman with CHD to avoid pregnancy?

Yes [ ] No [ ]

21. If your answer to Qs. 20 was 'Yes', which of the following cardiac lesions would you counsel women to avoid pregnancy with (please choose from the responses below)? *{You may choose more than one response}*

S/P Fontan palliations with single ***left*** ventricular physiology	[ ]
S/P Fontan palliations with single ***right*** ventricular physiology	[ ]
d-TGA S/P Mustard or Senning repair	[ ]
d-TGA S/P Arterial Switch Operation	[ ]
TOF s/p repair with residual moderate/severe pulmonary regurgitation	[ ]
Shunt lesions (unrepaired ASD's, VSD's)	[ ]
Aortic aneurysms ***with*** underlying connective tissue disease	[ ]
Aortic aneurysms ***without*** underlying connective tissue disease	[ ]
S/P Bio-prosthetic valve	[ ]
S/P Mechanical valve	[ ]

https://doi.org/10.1017/S104795111600038X Published online by Cambridge University Press

History of Pulmonary Hypertension [ ]

History of ventricular tachycardia (VT) [ ]

History of atrial arrhythmias [ ]

S/P Pacemaker [ ]

S/P ICD [ ]

Other (please specify):

_____

22. Have you recommended a therapeutic abortion to a woman with CHD ?

Yes [ ] No [ ]

If yes, please describe the underlying condition:

_____

23. Should women with CHD have their cardiology clinic visits (during pregnancy) at an adult hospital or children's institution?

Adult [ ] Pediatric [ ]

24. Do you feel that a high-risk obstetric team should deliver pregnant women with CHD ?

Yes [ ] No [ ]

25. Do you routinely discuss the care of your pregnant women with CHD with an obstetrician during their pregnancy?

Yes [ ] No [ ] I do not take care of pregnant women with CHD [ ]

26. Do you routinely discuss plans for labor/delivery or Caesarian section of your pregnant women with CHD with an anesthesiologist?

Yes [ ] No [ ] I do not take care of pregnant women with CHD [ ]

Thank you for your time.

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