



Guidelines for the Management of Adults with Congenital Heart Disease

Highlights for Patients and Families

- If you were told you had a congenital heart defect (CHD) as a child or adult, you should have cardiac follow up at an adult congenital heart disease (ACHD) program. You should see a board certified ACHD doctor even if you had surgery for your heart and were told that you were fixed. The ACHD medical team will check your heart function and make sure any surgeries or interventions you had are still working properly.
- If you are a teenager or a young adult with CHD, you should talk with your healthcare provider about transitioning and transferring to the care of a board certified ACHD doctor in an ACHD program. Not having knowledge about lifelong care for CHD results in gaps in care. In turn, this results in more hospital visits, a greater need for critical intervention, and more CHD-related issues.
- CHD heart problems are very different from “regular” adult-acquired heart problems. Approaches to diagnosis and treatment are **not** the same as they are in those adults with similar problems who do not

The American College of Cardiology and the American Heart Association made rules (guidelines) for taking care of adult patients with congenital heart defects (CHD). The first guidelines were released in 2008. Revised guidelines were released in 2018.

These guidelines give doctors the most important information for caring for adults with congenital heart disease (ACHD). They say it is important to be seen in ACHD programs and by ACHD board certified cardiologists. This summary includes recommendations based on the 2018 updated guidelines and will help you play an active role in the care of your heart. You can find the complete guidelines online at <https://bit.ly/2qS5e8j> and a summary of main points at <https://bit.ly/2YO6XrL>.

have CHD. All of the doctors, nurses, and other staff at your ACHD program should be trained in adult CHD. **Patients who are managed in ACHD programs generally have better outcomes and lower mortality than those who are not cared for in ACHD programs.**

- If you do not live near an ACHD program, you might need to travel to visit one. Your local heart team should talk to your ACHD medical team about your care. All of your doctors should check with your ACHD medical

team about any medications or treatments. In most cases, you should have all medical procedures at hospitals with an ACHD program.

- If you have complex CHD, the following should be done only by your ACHD program:
 - Echocardiograms
 - Pacemaker/ICD implantation
 - Cardiac MRI
 - Catheterizations – diagnostic and interventional
 - Cardiac CT/MUGA
 - Cardiac surgery

- Your ACHD board certified heart doctor will tell you how often you need to be seen. This will be based on your type of defect and how well your heart is functioning. You may have different long-term outcomes and/or problems than someone else with the same defect.
- Ask your ACHD medical team to teach you the name(s) of your CHD and any surgeries you have had. Know what you may be at risk for and the signs of new heart problems in the future.
- Your ACHD medical team should ask you about your emotional and social needs. They should offer support such as mental health support and social services like insurance and employment limitations. Mental health and neurodevelopmental issues are common in patients with ACHD. This includes anxiety, depression and post-traumatic stress disorder (PTSD). Dealing with any of these can affect your quality of life. It is important for your ACHD medical team to address them.
- Get an exercise prescription from your ACHD medical team and have it updated regularly. It is important that you live a heart-healthy lifestyle to prevent you from developing acquired heart disease. Exercising every day is important. This will help you strengthen your heart and maintain a healthy weight.
- If you receive emergency care, let your ACHD team know. Even if the emergency is not related to your heart, all emergency care providers need to talk with your ACHD medical team. Your ACHD medical team can decide if it is safe for you to continue your care locally or if you should be transferred to an ACHD program.
- Certain forms of birth control can be risky for women with CHD. You should talk to your ACHD medical team to find a choice that is safe and effective for you.
- You should get pre-pregnancy counseling from your ACHD medical team **before** becoming pregnant. With the right care, most women with CHD can have a baby. The ACHD medical team should review the risks of pregnancy to both you and your baby during pregnancy, delivery and the postpartum period. They should also talk about the potential long term risks to the mother. In some cases, more heart treatment or adjusting medication might need to occur before you become pregnant.
- If you are experiencing infertility, talk with your ACHD medical team. Counseling should include the risks and benefits of alternative methods of having a family. Ask for a referral to a reproductive endocrinologist if needed.
- As sexuality is an important aspect of your quality of life, it is important to address any concerns with your ACHD medical team.
- ACHD patients are at higher risk of having an infection of the heart valves, or infective endocarditis. You should talk to your ACHD medical team to see if you need to take preventive antibiotics before dental work and certain procedures.
- If you are an adult with CHD who also has risk factors for acquired heart disease, you should talk to your ACHD medical team about an evaluation. Prevention and treatment of conditions that can lead to acquired heart disease are important. This includes promoting a healthy lifestyle in all patients with ACHD.
- If you received blood transfusions during surgery before 1992, you might be at risk for hepatitis C. If you had surgery before then, you should ask your ACHD medical team to screen you for hepatitis C.
- Before having non-cardiac surgery, you should consult with your ACHD medical team. You may be at a higher surgical risk than patients who do not have CHD. Complex ACHD patients and/or those with progressive disease should receive evaluation and any surgery or other intervention in collaboration with an ACHD program.

To learn more about the 2018 ACHD Guidelines and putting them into practice for your ACHD care, visit the following links to view webinars given by our expert Medical Advisory Board members:

- What You Need To Know: <http://bit.ly/ACHDPart1>
- Take Home Messages: <http://bit.ly/ACHDPart2>