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EDITORIAL COMMENT

New Findings on Ascending Aortic Dilation in Coarctation of the Aorta

Expanding Perspectives*

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With the advent of modern imaging techniques and specialized cardiovascular surgery, 90% to 97% of children with congenital heart disease are now expected to reach adulthood.[1] Although this is a major triumph for modern pediatric cardiovascular care, it also means that the prevalence of adult congenital heart disease is continuously rising, necessitating expanded research on long-term complications and care in this unique patient group.

One such study is presented in this issue of the *Journal of the American College of Cardiology*. Performed by Egbe et al,[2] it provides an extensive look at the relationship among coarctation of the aorta (COA), bicuspid aortic valve (BAV), and ascending aortic dilation. It has long been known that these conditions are related, but the exact causality has remained somewhat elusive until now.

COA was one of the first congenital heart lesions to be successfully repaired. Unfortunately, repaired does not necessarily mean cured. As in many other forms of congenital heart disease, studies have shown that even after the structural anomaly is mended, patients born with COA are still at risk of several, often severe complications.[3,4]

The prevalence of arterial hypertension is high in adults with repaired COA.[5,6] The reasons are likely multifactorial, with dysfunction of blood pressure regulation, stiff segments in the descending aorta, and endothelial dysfunction having all been suggested as contributing factors. As the left ventricle is exposed to an increased pressure load, these patients are consequently also at risk of developing left ventricular hypertrophy.[7]

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Furthermore, recurrent narrowing at the site of the previous intervention may occur, and aneurysms may develop in both the ascending aorta and at the site of repair, especially if synthetic prosthetic material has been used. Further complicating the issue, approximately 80% of patients with COA also have BAV.[8-10] BAV is known to be associated with the development of aortic stenosis, aortic regurgitation, and aneurysms in the ascending aorta.[11-13] In fact, the majority of patients with BAV will develop significant aortic valve disease and/or aneurysm of the ascending aorta at some point in their life. Also, a recent study showed that 29% of patients with an index operation for COA needed at least 1 additional intervention on the aortic valve, left outflow tract, the coarctation site, or the ascending aorta.[14] All of these possible complications clearly demonstrate the need for systematic follow-up of patients with repaired COA.

Egbe et al[2] present a large retrospective analysis of ascending aortic dilation in patients with and without COA and BAV, respectively. The 867 COA patients included in the study were matched with 867 patients with isolated BAV. Similarly, they were also matched with a control group of 867 individuals without any structural heart disease.

The main takeaway from this robust study is that a patient with COA and a tricuspid aortic valve was no more likely to develop an ascending aortic aneurysm

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than a person without any structural heart disease. The risk of developing an ascending aortic aneurysm was instead dependent on the presence of BAV, whether or not the patient was born with COA. Additional findings showed that the risk of acute aortic complications was low in all groups, and that arterial hypertension was strongly associated with progressive dilation of the ascending aorta.

In cardiovascular medicine, we are used to treatments and guidelines being based on large, prospective, randomized trials. Although there is ample data regarding major fields such as ischemic heart disease, heart failure, and various device treatments, when it comes to adult congenital heart disease, randomized trials are sparse and guidelines are therefore often based on expert opinion and previous experience. This makes studies like the one conducted by Egbe et al[2] very welcome in our field. However, one should bear in mind these studies' retrospective nature and single-center recruitment of patients, the latter of which may introduce a bias, as patients are often treated in a dedicated milieu with the benefits of experienced staff and high patient volumes.

The clinical implications of the study mainly concern the follow-up of patients with COA. While the risk of dilation or recoarctation near the site of repair still necessitates imaging surveillance, the study demonstrates that COA does not, in itself, cause an increased risk of aortic dilation in patients with a tricuspid aortic valve. Also, it showed that the risk of dissection and other acute aortic complications is low. Despite the single-center design, it is reasonable to assume that these conclusions are generally applicable when patients are in appropriate follow-up care, and when the current recommendations for prophylactic interventions are applied. However, it is possible that the outcomes may not be quite as favorable in less specialized contexts, but, if so, the results of this study still provide a goal to strive toward.

In addition, the findings once again demonstrate the need for adequate blood pressure regulation in patients with repaired COA given that arterial hypertension was related to progressively increasing aortic diameter. Because these patients tend to be younger when arterial hypertension manifests, even a slightly increased blood pressure may act upon the heart and vessels for a long period of time. Therefore, optimal blood pressure for this patient group may differ from the recommended limits for hypertensive patients with no underlying structural heart disease, at least from a long-term perspective.

In the future, it would be preferable to base the treatment of arterial hypertension in COA on large-scale prospective trials focused on this specific patient group. Hopefully, the time will soon come for international collaborative efforts in this field, further optimizing the therapeutic options for patients born with heart defects.

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REFERENCES

1. Mandalenakis Z, Giang KW, Eriksson P, et al. Survival in children with congenital heart disease: have we reached a peak at 97. *J Am Heart Assoc*. 2020;9:e017704.

2. Egbe AC, Miranda WR, Abozied O, et al. Coarctation of aorta with tricuspid aortic valve is not associated with ascending aortic aneurysm. *J Am Coll Cardiol*. 2024;83(12):1136–1146.

3. Toro-Salazar OH, Steinberger J, Thomas W, Rocchini AP, Carpenter B, Moller JH. Long-term follow-up of patients after coarctation of the aorta repair. *Am J Cardiol*. 2002;89:541–547.

4. Celermajer DS, Greaves K. Survivors of coarctation repair: fixed but not cured. *Heart*. 2002;88:113–114.

5. Rinnström D, Dellborg M, Thilen U, et al. Hypertension in adults with repaired coarctation of the aorta. *Am Heart J*. 2016;181:10–15.

6. Rinnström D, Dellborg M, Thilen U, et al. Poor blood pressure control in adults with repaired coarctation of the aorta and hypertension: a register-based study of associated factors. *Cardiol Young*. 2017;27:1708–1715.

7. Rinnström D, Dellborg M, Thilen U, et al. Left ventricular hypertrophy in adults with previous repair of coarctation of the aorta; association with systolic blood pressure in the high normal range. *Int J Cardiol*. 2016;218:59–64.

8. Teo LL, Cannell T, Babu-Narayan SV, Hughes M, Mohiaddin RH. Prevalence of associated cardiovascular abnormalities in 500 patients with aortic coarctation referred for cardiovascular magnetic resonance imaging to a tertiary center. *Pediatr Cardiol*. 2011;32:1120–1127.

9. Jurcut R, Daraban AM, Lorber A, et al. Coarctation of the aorta in adults: what is the best treatment? Case report and literature review. *J Med Life*. 2011;4:189–195.

10. Rinnström D, Engstrom KG, Johansson B. Subtypes of bicuspid aortic valves in coarctation of the aorta. *Heart Vessels*. 2014;29:354–363.

11. Ward C. Clinical significance of the bicuspid aortic valve. *Heart*. 2000;83:81–85.

12. Tzemos N, Therrien J, Yip J, et al. Outcomes in adults with bicuspid aortic valves. *JAMA*. 2008;300:1317–1325.

13. Tadros TM, Klein MD, Shapira OM. Ascending aortic dilatation associated with bicuspid aortic valve: pathophysiology, molecular biology, and clinical implications. *Circulation*. 2009;119:880–890.

14. Meidell Blylod V, Rinnström D, Pennlert J, et al. Interventions in adults with repaired coarctation of the aorta. *J Am Heart Assoc*. 2022;11:e023954.

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