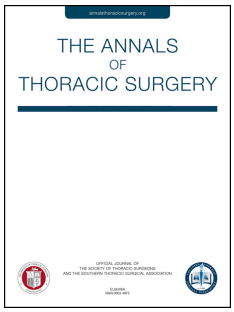

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Adult Congenital Heart Disease Surgery: On Track to Succeed or Time to Reset?

Running Head: ACHD Surgery: On Track or Reset Time?

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MANAGEMENT OF ACHD PATIENTS — SCOPE OF THE PROBLEM

Congenital heart disease requires lifelong maintenance with the help of a specialized team and regular subspecialized care has been independently associated with decreased mortality. ACHD patients have higher risk of mortality due to non-CHD related illnesses than age-matched peers, as these patients have underlying heart defects which increases morbidity and mortality related to non-CHD health issues (1). Mortality and morbidity rates amongst the ACHD population are not solely attributed to their underlying cardiac defect but are also from reported gaps in care such as accessibility, provider apprehension, and patients' lack of knowledge of their disease (2). Difficulties in navigating changes or loss of insurance, distance to/from clinical locations, and fewer trained team members being some of the most frequently reported reasons for loss to follow-up (2). Patients also report that their provider told them follow-up was not necessary, and nearly a third, cannot name their actual heart defect (2).

A once solely pediatric disease evolving into an adult subspecialty is not unique to CHD. With the advancement of diagnostic technology, reparative procedures, and treatments, many congenital diseases now have their own adult treatment centers, fellowships, and recognized societies. In 1984, Surgeon General Dr. Everett committed that youth and adolescents aging into adulthood are promised commitment to improved services, trainings, and research (3). In 1993, the Society for Adolescent Medicine emphasized the need for the transition of chronically ill and disabled adolescents to adult oriented programs (4). In 2002, a consensus statement on healthcare transition needs was approved as policy by the four medical boards: American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians-American Society of Internal Medicine (5). These policies and efforts lay out acceptable guidelines for patients with chronic healthcare issues transitioning from pediatric to adult care and include development of appropriate healthcare services (5).

Similar to most chronic medical conditions, transition from pediatric congenital heart care to adult care has the highest instance of loss to follow-up with a reported average gap of 22 years (6). These gaps are a significant risk factor for premature mortality in ACHD patients. Patients without follow up are three times more likely to require immediate cardiac intervention on presentation, and half of the deaths occur when patients were not cared for by specialized care teams. Despite these findings 42% of ACHD patients report a gap in care and nearly a quarter never establish care at a specialized ACHD clinic. Successful transition to adult programs are most successful when aided by a patient's own CHD pediatrician or with a formal transition program.

Pediatricians also contribute to the gaps in care for their patients as a result of their lack of trust in the adult services/systems receiving their patients. Pediatricians have reported apprehension about and significant delays in, transitioning their patients to adult care, primarily as a result of a lack of confidence in available and qualified ACHD providers. Patients have also reported that a lack of current healthcare provider knowledge regarding CHD has contributed to gaps in care (2). Pediatric cardiologists self-report a decline in competence from 94% in treating patients under the age of 21, to 53% once patients reach the age of 30 (7). Among adult patients who do get transferred, 80% are referred to adult programs due to adult comorbidities.

Currently, there are fewer than 50% of the ACHD centers predicted to be required to support the estimated ACHD patient population. In order to determine the number of specialty clinics necessary to care for the ACHD population, Marelli and colleagues created a model based on the 2000 Bethesda recommendations — they determined that there would need to be 1 clinic per 2 million adults with an assumed ACHD prevalence of 4.4/1000 people (8). By

adjusting the model to reflect the Gilboa prevalence of 6.16/1000, the number of ACHD clinics needed would be 1 per 940,000 adults (9). By using a forecast model and assuming a constant growth rate of established ACHD centers, a projected estimate of needed centers shows a major deficit. These values likely understate the gap, as the evidence is clear that the ACHD population is growing at a rapid rate and reflects a crucial need to increase access and these need to be populated by subspecialists with the necessary medical and surgical armamentarium (Figure 1).

SURGERY IN ACHD PATIENTS — CONSIDERATIONS

In many healthcare systems, systematic care of surgical ACHD patients has lagged compared to pediatric CHD patients. With the upcoming addition of the ACHD module to the Society of Thoracic Surgeons Congenital Heart Surgery Database we will be able to begin to document the scope of the care needed by this cohort. We know that more than 20% of ACHD patients had at least one operation in adulthood, many of which were first time operations (10). The chance of needing an operation increases with age, and patients over 60 have a 43% chance of needing an operation related to their heart defect. In less than 10 years, the median age of ACHD patients had increased from 53 years to 58 years, and over 20 years, the number of ACHD patients over the age of 60 presenting to ACHD clinics has increased by 8-fold (11). The surgical caseload — epicardial leads, abdominal pacemaker procedures, endocarditis, coronary revascularization in redo ACHD setting, acquired valve disease, aortopathy, conduit changes, anomalous coronary procedures, transcatheter valve or stent implantation, heart/lung transplant, and mechanical circulatory support for heart failure to name a few — will continue to increase as the median age of the ACHD population advances.

ACHD cases are often surgically complex, however despite this there are data that suggest the overall mortality rate for adult congenital cases should be ≤2% (12) The current congenital heart surgery database does not address contemporary complexity nor risk, and therefore these rates are deduced from published series that do not reflect the reality, further supported by the fact that none of ACHD surgeries exceed STAT 3 category. Studies have shown that like pediatric CHD surgical cases, nearly half of ACHD surgeries have at least one complication.

Social determinants of health are pertinent to medical histories in adult patients and need to considered. Patients who wish to become pregnant are at increased risk of pregnancy-related complications compared to age-matched controls. Hormonal fluctuations increase the risk of thrombosis and many common cardiac medications are known teratogens. A past medical history of blood transfusion or other cardiovascular diseases increase the risk of surgical complication for the patient. Patients with significant medical histories often face longer wait times for transplants and increased operation time which further compounds the risk of complication occurrence. These factors highlight the importance of surgeon and programmatic experience and sensitivity to comorbidities and social factors.

As mentioned, inadequate pediatric cardiac surgical resources may result in patients being treated at centers by subspecialists who are not qualified enough and with low ACHD surgical case volume. For example, if there is no ACHA accredited center where patients can be treated by congenitally trained surgeons who are experienced in ACHD, patients may be referred to the closest available adult cardiac surgeon. This type of migration results in suboptimal care for the individual patient as well as in reduced numbers of ACHD cases at designated specialty centers (a “lose-lose” situation) (13). There has been a well-documented relationship between volumes and outcomes in pediatric CHD surgery. Hospitals that manage

higher case volumes in general, have better outcomes. This is in part attributed to increased surgeon experience and expertise in handling cases. There is an inverse relationship between surgical mortality and case volume which has been reported in pediatric CHD surgery outcomes (14). Completely apart from institutional experience and surgeon ability, patient and management factors have been identified as being key in outcomes in neonates undergoing congenital heart surgery (15). Mortality rates start increasing when there are fewer than 60 cardiopulmonary bypass CHD surgical cases performed annually, and in Europe, an optimal structure of a congenital heart surgery program has been articulated as including at least 3 congenitally trained surgeons who perform minimum of 125 cases per year each (16).

There is a marked discrepancy between ACHD surgical volume and the guidelines articulated for pediatric CHD and adult cardiac surgery. The current report of mean annual volume of six ACHD CPB cases among adult cardiac surgeons who perform such operations is a very low number regardless of what bar is used (17). The ACHD caseload for congenitally trained surgeons is not much better. A 2015 survey estimated that in 297 active congenital heart surgeons, a third performed fewer than 25 ACHD surgeries annually and only 12% performing more than 50 annual ACHD surgeries (13). Why are there standards in place for the pediatric CHD population and the non-CHD adult cardiac patients, but not for ACHD patients who arguably have some of the most complex repeat interventions? Between the congenitally trained surgeon whose practice mainly focuses on pediatric cases, and the adult cardiac surgeon who mainly practices on non-congenital heart disease, the ACHD patient may be orphaned, without anywhere to call home.

Operations performed by congenitally trained surgeons have resulted in better outcomes for the ACHD patient (17, 18). However, having congenitally trained surgeons take on all ACHD cases is not feasible. Even the small percentage of ACHD cases that are handled in pediatric

specialties creates economic burdens. ACHD patients are associated with high resource use and can cost pediatric departments $1B annually (19). Surveys show that of the estimated 300 practicing congenital surgeons, a third expected to retire within 10 years (13). The surgical workforce among non-congenital cardiac surgeons is also a cause for concern. If we accept that there are not enough pediatric cardiac surgeons to provide care for ACHD patients now and into the future, then we will be depending on adult cardiac surgeons with additional training to provide some of this care. However, like pediatric cardiac surgeons, there is a predicted upcoming shortfall of adult cardiac surgeons. Based on current projections, by 2030, there will be a net deficit of two thousand cardiac surgeons (20). Within the same time frame, projection models estimate that there will be a 60% increase in cardiac procedures which will create a near impossible caseload for the adult cardiac surgeon. Without an increase in funding and cardiothoracic surgical training the surgeon shortage will disproportionally affect an already underserved ACHD population.

CONCLUSION

The current approach to adult congenital heart surgery is unsustainable and is a substantial disservice. Training and certification in pediatric congenital heart surgery and medical management of ACHD has improved health outcomes and overall quality of care. Similar measures must be taken to address the surgical care of ACHD patients. Accordingly, comprehensive ACHD centers should be established and supported. Quality infrastructure around management of ACHD patients is required. As care for this patient population grows, anesthesia, critical care, perfusion, nursing, cardiology, interventional proceduralists and radiologists will continue to contribute and learn from each other. How do we make this happen in our current surgical education model, where do we structurally follow these patients, how do we construct a productive and professional mechanism to span the pediatric and adult

congenital surgeon while covering necessary clinical requirements in both frameworks of care will be important next steps in readiness to care for the ACHD population.

Figure Legend

Figure 1. ACHD Clinical Gap in US. The blue line estimates US population in millions, the green line shows ACHD prevalence in the US, and the grey line estimates the ACHD clinics needed to manage patients.

REFERENCES

1. Agarwal A, Thombley R, Broberg CS et al. Age- and lesion-related comorbidity burden among us adults with congenital heart disease: A population-based study. Journal of the American Heart Association 2019;8(20):e013450.

2. Gurvitz M, Valente AM, Broberg C et al. Prevalence and predictors of gaps in care among adult congenital heart disease patients: Heart-achd (the health, education, and access research trial). J Am Coll Cardiol 2013;61(21):2180-2184.

3. Koop C, Magrab P, Millar H. Surgeon general's conference: Growing up and getting medical care: Youth with special health care needs. A summary of conference proceedings. Jeckyll Island, Georgia 1989.

4. Newacheck PW, Taylor WR. Childhood chronic illness: Prevalence, severity, and impact. American Journal of Public Health 1992;82(3):364-371.

5. Pediatrics AAo, Physicians AAoF, Medicine ACoP-ASoI. A consensus statement on health care transitions for young adults with special health care needs. Pediatrics 2002;110(Supplement 3):1304-1306.

6. Wray J, Frigiola A, Bull C. Loss to specialist follow-up in congenital heart disease; out of sight, out of mind. Heart 2013;99(7):485-490.

7. Fernandes SM, Khairy P, Fishman L et al. Referral patterns and perceived barriers to adult congenital heart disease care: Results of a survey of u.S. Pediatric cardiologists. J Am Coll Cardiol 2012;60(23):2411-2418.

8. Marelli AJ, Therrien J, Mackie AS, Ionescu-Ittu R, Pilote L. Planning the specialized care of adult congenital heart disease patients: From numbers to guidelines; an epidemiologic approach. Am Heart J 2009;157(1):1-8.

9. Gilboa SM, Devine OJ, Kucik JE et al. Congenital heart defects in the united states: Estimating the magnitude of the affected population in 2010. Circulation 2016;134(2):101-109.

10. Zomer AC, Verheugt CL, Vaartjes I et al. Surgery in adults with congenital heart disease. Circulation 2011;124(20):2195-2201.

11. Lui GK, Saidi A, Bhatt AB et al. Diagnosis and management of noncardiac complications in adults with congenital heart disease: A scientific statement from the american heart association. Circulation 2017;136(20).

12. Jain PN, Salciccioli KB, Guffey D et al. Risk factors for perioperative morbidity in adults undergoing cardiac surgery at children's hospitals. Ann Thorac Surg 2022;113(6):2062-2070.

13. Morales DLS, Khan MS, Turek JW et al. Report of the 2015 society of thoracic surgeons congenital heart surgery practice survey. The Annals of thoracic surgery 2017;103(2):622-628.

14. Welke KF, O'Brien SM, Peterson ED, Ungerleider RM, Jacobs ML, Jacobs JP. The complex relationship between pediatric cardiac surgical case volumes and mortality rates in a national clinical database. The Journal of Thoracic and Cardiovascular Surgery 2009;137(5):1133-1140.

15. Karamlou T, McCrindle BW, Blackstone EH et al. Lesion-specific outcomes in neonates undergoing congenital heart surgery are related predominantly to patient and management factors rather than institution or surgeon experience: A congenital heart surgeons society study. J Thorac Cardiovasc Surg 2010;139(3):569-577 e561.

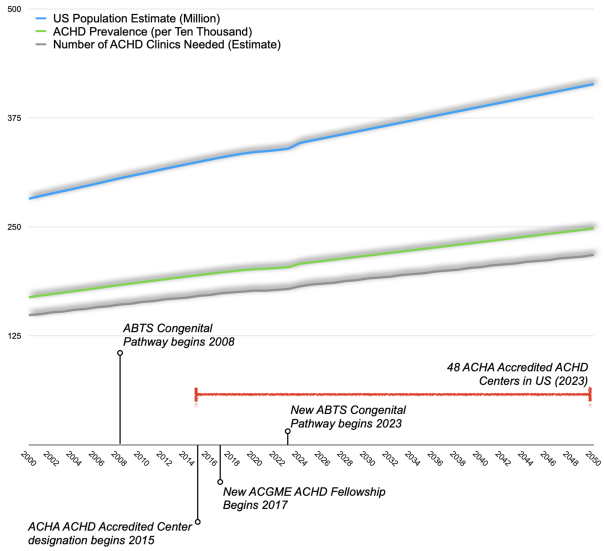
16. Daenen W, Lacour-Gayet F, Aberg T et al. Optimal structure of a congenital heart surgery department in europe: By eacts congenital heart disease committee1. Elsevier Science BV;2003.

17. Karamlou T, Diggs BS, Person T, Ungerleider RM, Welke KF. National practice patterns for management of adult congenital heart disease. Circulation 2008;118(23):2345-2352.

18. Guleserian KJ. Adult congenital heart disease: Surgical advances and options. Progress in Cardiovascular Diseases 2011;53(4):254-264.

19. Kim YY, Gauvreau K, Bacha EA, Landzberg MJ, Benavidez OJ. Resource use among adult congenital heart surgery admissions in pediatric hospitals. Circulation: Cardiovascular Quality and Outcomes 2011;4(6):634-639.

20. Williams WG. Congenital heart disease: Interrelation between german diagnoses-related groups system and aristotle complexity score. Eur J Cardiothorac Surg 2010;37(6):1276-1277.



Declaration of interests

☐ The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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