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A Comprehensive Insight into Adult Congenital Heart Disease: A Battle of Survival into Adulthood

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Abstract: Adult Congenital Heart Disease (ACHD) is an anatomical anomaly of the heart or the great vessels. Today, ACHD, according to the 2018 AHA guidelines and 2020 ESC guidelines, corresponds to its anatomical and physiological flaws. Along with these congenital lesions, ACHD can further be combined with Eisenmenger syndrome, backed up by Pulmonary Hypertension. Apart from these, existing comorbidities like Heart Failure or structural valvular issues in ACHD patients add to the numerous challenges faced to survive into adulthood. Aiding clinicians in diagnosing ACHD is Echocardiography, Heart CT angiography and Heart MRI. Usually, other differential diagnoses must be eliminated to establish a confirmed diagnosis of ACHD, following a detailed patient and family history, backed up by genetic testing. Based on each patient's unique anatomy, clinical condition, and long-term health objectives, the care of ACHD is a multifaceted and protracted procedure that includes preventive, surgical and interventional techniques, rhythm management, and psychosocial support. Gradually changing its emphasis from primary surgery to the careful management of acquired comorbidities, late complications, and residual abnormalities. Limited anatomical defect care is giving way to an era of advancements marked by proactive and individualised strategies. Ongoing research is enabling innovations that integrate regenerative medicine, artificial intelligence, imaging, and new and improved devices. This paper reviews the essential details of ACHD and stresses the importance of advanced diagnostic modalities and associated therapy for better care and survival of patients with ACHD.

Keywords: *Adult Congenital Heart Disease, Congenital Heart Disease, Cyanotic Heart Lesions, Acyanotic Heart Lesions, Eisenmenger Syndrome.*

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I. INTRODUCTION

Congenital heart disease (CHD) is a significant anatomical anomaly of the major intrathoracic arteries or the cardiac muscle, possessing practical or possible physiological consequences [1]. The area of ACHD is expanding significantly as a result of the growing patient base [2]. The majority of people with CHD who are still alive have made it to adulthood, and they now outnumber newborns who were born with this disease [3]. Although CHD was once a condition mainly found in specialised pediatric facilities, technological advancements have significantly raised the total number of people who survive into adulthood, despite the number of challenges faced in non-specialised emergency rooms [4]. Among adolescents with CHD, arrhythmias are among the most frequent causes of hospitalisation among all lesion stages and are factors responsible for excessive fatality in this demographic group [5]. Various diagnostic modalities like Echocardiography, Heart CT angiography and MRI play a significant role in contributing towards the development of ACHD therapy in adulthood.

Eventually, the majority of patients remain uncured despite advancements in diagnosis, the application of devices and interventional treatments, and increased survival [1]. They pose certain obstacles that demand specialised

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treatment and an integrated approach [2]. Since timely rhythm management, pulmonary vasodilators, endocarditis prevention, and re-intervention can have significant effects on survival, it is crucial to comprehend their courses and identify these complicated issues early. Although there is a substantial care gap, because many general cardiologists lack the necessary training to handle the complicated and ongoing care requirements of patients with ACHD [3], thus concluding that the treatment of such patients must include endocarditis specialists [6]. However, when alternative treatments fail and the patient is classed as severely fragile, the majority of CHD patients are evaluated for organ transplantation. As a result of technical difficulties or a lack of resilience, these patients either pass away while awaiting organs or soon after transplantation. Therefore, determining clinical indicators that might forecast the best timing for transplantation in individuals with ACHD is crucial [7]. This narrative focuses on identifying not only the clinical factors of ACHD, but also the classification, various long-term complications, diagnostic measures, managing ACHD, and areas of development in the future.

II. ANATOMICAL AND PHYSIOLOGICAL CLASSIFICATION OF ACHD.

The most notable improvement in adult survival in CHD patients is seen in those with the most severe condition; 90% of children with severe congenital heart disease are now anticipated to survive to age 18 [8]. A comprehensive classification system that integrates anatomical features and physiological effects is necessary for the management of ACHD [9]. Various guidelines, like the 2020 ESC guidelines, primarily classify ACHD based on its anatomic abnormalities into Mild, Moderate and Severe [10], while the revised 2018 AHA guidelines stratify ACHD based on anatomy as simple, moderate complexity and severe complexity as shown in Table 1, as well as on physiology, graded from Stage A to stage D demonstrated in Table 2, for better management of patients [8]. In previous archives such as the 2008 ACHD guidelines, the classification was solely based on anatomy and used to assess the severity of the condition, patients with the same underlying architecture undergo quite distinct repairs and experience diverse physiological effects, as seen in a patient of the same age with tetralogy of Fallot (TOF) may have undergone palliative shunting followed by a transannular patch repair, which resulted in severe pulmonary regurgitation (PR) with right ventricular (RV) enlargement, biventricular dysfunction, and ventricular tachycardia (VT), while some patients may have excellent biventricular function, normal exercise capacity, and no arrhythmias following a primary valve-sparing repair[8]. Hence, the writing committee of AHA created the ACHD Anatomic and Physiological (AP) classification system to more thoroughly classify the disease severity in CHD [11].

Table 1 Comparison Between the 2018 AHA and the 2020 ESC Guidelines for ACHD.

	Anatomical classification according to AHA guidelines	**Anatomical classification according to ESC guidelines**	**References**
Simple /Mild	● Localised ASD ● Localised VSD ● Minimally diffused pulmonic stenosis	● ASD, VSD, PDA ● Localised aortic and mitral valve disease ● Mild pulmonic stenosis	[8,10]
Moderate complexity	● Partial or total AVSD ● Hereditary aortic and mitral valve diseases, ● Aortic coarctation, ● Ebstein anomaly ● Corrected ToF	● TGA after the switch operation ● Cardiac manifestations of Marfan and Turner syndromes ● Twin-chambered right ventricle	[8,10]
Complex/Severe	● Twin-outlet ventricle ● Disruption of the aortic arch ● Ventricular inversion ● Criss-cross hearts	● Fontan procedure ● TGA patients without atrial switch surgery ● Univentricular heart ● Any type of CHD associated with pulmonary valve diseases, like Eisenmenger syndrome	[8,10]

Abbreviations: AHA: American Heart Association; ESC: European Society of Cardiology; ACHD: Adult Congenital Heart Disease; ASD: Atrial Septal Defect; Ventricular Septal Defect; PDA: Patent Ductus Arteriosus; AVSD: Atrio-Ventricular Septal Defect; TGA: Transposition of Great Arteries; CHD: Congenital Heart Disease.

As mentioned earlier, AHA guidelines classify ACHD as simple, moderate and complex lesions based on its anatomy. Simple lesions consist of localised minor ASD and VSD, minimally diffused pulmonic stenosis and other repaired lesions like the Ductus arteriosus that was formerly blocked or fused, rectified sinus venosus defect or secundum ASD without a sizable residual shunt or chamber expansion and corrected VSD without appreciable chamber enlargement or residual shunt [8].

The category of moderate lesions includes a fistula between the aorta and the left ventricle, a partial or complete abnormal pulmonary venous connection, or the pulmonary artery giving rise to an abnormal coronary artery. It also includes an anomalous coronary artery aortic origin from the opposing sinus, partial or total AVSD, which also incorporates primum ASD, hereditary aortic and mitral valve diseases, and aortic coarctation or constriction. Other lesions, like mild, moderate and severe variations in the disease

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spectrum of Ebstein anomaly, blockage of the right ventricle's infundibular outflow, ASD in Ostium Primum, unrepaired secundum ASD that is moderate and large, large, moderate and consistent patent ductus arteriosus, moderate or severe regurgitation and stenosis of the pulmonary valve, aneurysm or fistula of the sinus of Valsalva, defect of the sinus venosus, subvalvar and supravalvular aortic stenosis, Atrioventricular valve straddling, repaired tetralogy of Fallot and VSD with concomitant abnormalities and/or moderate or larger shunts, also fall under moderate lesions of AHA classification [8].

Whereas the great complexity or complex lesions include palliated or unrepaired congenital cardiac defects that are cyanotic, a ventricle with two outlets, the Fontan method, which is performed on children with single-ventricle heart abnormalities who have palliative open heart surgery. It also includes disruption of the aortic arch and absence of the left atrioventricular junction, called mitral atresia. It also comprises any anatomical anomaly that results in a functionally single ventricle, such as tricuspid atresia, hypoplastic left heart, or twin inlet left ventricle, all types of pulmonary atresia, TGA, and common atrial trunk, usually referred to as truncus arteriosus. Additional atrioventricular and ventriculo-arterial connection anomalies, such as ventricular inversion, heterotaxy syndromes, isomerism, and crisscross hearts, are also inculcated under the severe complexity or complex lesions of ACHD [8].

In contrast, the 2020 ESC guidelines classify ACHD as mild, moderate and severe as stated previously [9].

Mild lesions consist not only of ASD, VSD, pulmonic stenosis and repaired sinus venosus defect, VSD, PDA, or secundum ASD without any aftereffects, including ventricular failure, increased PAP, or chamber enlargement, similar to the simple lesions of AHA classification, but also include PDA, localised inherited Aortic and Mitral valve diseases as well as bicuspid aortic disease [9].

While the majority of moderate lesions are similar in the two guidelines, twin-chambered right ventricle, cardiac manifestations like aortic aneurysm and subsequent dissection are the most frequent and dreaded consequences, along with arrhythmia and cardiomyopathy, which are potentially fatal, are seen in Marfan syndrome, making them the leading cause of mortality [12]. Absence of the left main coronary artery, cyanotic heart disease, or Hypoplastic left Heart syndrome, PDA and dextrocardia seen in Turner syndrome [13], as well as Transposition of the great arteries after the atrial switch surgery, all fall under the moderate lesions of the 2020 ESC guidelines, setting it apart [10].

The Severe lesions category of ESC guidelines comprises the same type of lesions as the complex lesions of the AHA classification system, like palliated or non-corrected forms of hereditary congenital heart defects, Twin-outlet ventricle, the Fontan circulation, pulmonary and mitral atresia. Although the ESC guidelines include patients with TGA after atrial switch surgery under the moderate lesions category, as compared to the AHA guidelines, which include the TGA lesion, whether classic or d-TGA and Congenitally corrected TGA under the complex lesions category. Any kind of CHD associated with Pulmonary valve disease, like the Eisenmenger syndrome, also falls under the severe lesions category of the ESC guidelines [10]. Elevated pulmonary vascular resistance and right-to-left blood shunting via a systemic-to-pulmonary circulation link are hallmarks of Eisenmenger syndrome, while the majority of individuals with the illness live up to 20-30 years. The hyperviscosity syndrome, cerebrovascular problems, or thromboembolic events can result from the hemostatic alterations linked to this syndrome [14].

The AHA also stratifies ACHD based on its physiologic effects, from stage A to stage D, in correlation with the New York Heart Association Functional Classification [NYHA FC] guidelines for HF. Table 2 demonstrates the physiological classification system of ACHD.

Table 2 Physiological Classification of ACHD According to the 2018 AHA Guidelines and in Correlation with the NYHA FC Guidelines for Heart Failure

AHA classification stages of ACHD	Physiologic effects	NHYA FC symptoms	References
Stage A	● Absence of anatomical and hemodynamic after effects ● Absence of arrhythmias ● No changes in renal/pulmonary/hepatic function	Class I: No anatomical or hemodynamic after effects No irregular heartbeat	[8,15]
Stage B	● mild ventricular dysfunction, ● mild aortic enlargement, ● mild ventricular enlargement ● Valvular disease that is mild ● minor or trivial shunt ● Arrhythmia not needing medical intervention	Class II: some restriction on exercise. Regular exercise causes chest pain, palpitations, exhaustion, and shortness of breath.	[8,15]
Stage C	● Remarkable valvular disease ● Modest aortic enlargement ● Mild to moderate hypoxemia ● Arterial or venous stenosis ● Medically controlled therapy ● End-organ malfunction, which is treatable	Class III: noticeable restriction in physical activity. Fatigue, palpitations, dyspnea, or chest pain are experienced.	[8,15]

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Stage D	• extreme expansion of the aorta • Treatment-resistant arrhythmias • Acute hypoxemia, which is usually linked to cyanosis • profound pulmonary hypertension • The Eisenmenger disease • Therapy-resistant End-organ malfunction	Class IV: signs of resting heart failure. Any physical exertion exacerbates the pain.	[8,15]

Similar to the American Heart Association's (AHA) Heart Failure, A - D classification, which is frequently used to categorise the population with ACHD, [16]. Compared to the AHA Heart Failure scale superimposed over various anatomic variants, this new categorisation is anticipated to more accurately characterise the current state of the disease and takes into consideration other physiological issues that are frequently encountered by ACHD patients. However, the classification aims to standardise management and assessment.

Its capacity to accurately determine the overall severity of the disease will need to be evaluated over time. The classification method should facilitate data gathering and, eventually, offer chances to critically assess its effectiveness with reduced heterogeneity [16]. In contrast, the ESC guidelines offer to meet the needs of ACHD patients with specific organisational and structural healthcare requirements. However, according to ESC guidelines, congenital heart abnormalities are arbitrary classifications of mild, moderate, and severe lesions. It is yet to be determined whether this classification is useful for risk stratification and clinical management in sizable registries [10].

III. LONG-TERM COMPLICATIONS

While congenital heart surgery has advanced, most ACHD patients have a lifelong course of progressing problems. Anatomic corrections are not physiologic cures and over time, residual lesions, hemodynamic stresses, residual dysplastic vessels, and previous interventions may lead to progressive dysfunction. Heart failure takes its place as one of the prime long-term concerns for ACHD. This is particularly true for those with residual shunts or valve lesions, which subject the heart to continuous pressure or volume overload. With time, this strain contributes to the gradual weakening of the ventricular walls, especially the right ventricle (RV). In individuals who have undergone repair for ToF, RV dysfunction remains a key contributor to poor long-term outcomes [17]. Furthermore, RV outflow variations are nearly always present following ToF repair [18].

IV. DIAGNOSTIC MODALITIES

A range of new challenges, a comprehensive, multimodal approach is now more important as survival into adulthood with CHD is on the rise. This evaluation process commences with a methodical clinical history that records surgical and catheter interventions, the progression of symptoms, and any complications that may have been encountered. Following this, a physical examination is conducted to identify signs such as cyanosis, heart murmurs, indications of heart failure, and circulatory congestion. Basic diagnostic tests like ECG, to help detect arrhythmias and myocardial infarction along with chest X-ray to help identify chamber enlargement, conduction issues, cardiomegaly, altered pulmonary vascularity, surgical clips, and heart failure signs, providing essential clinical context before advanced imaging [27].

This interaction between mechanical and electrical stress contributes to the relatively elevated occurrence of arrhythmias in individuals with ACHD. The surgical scars and dilated chambers form an ideal substrate for atrial flutter, atrial fibrillation (AF), and ventricular tachycardia (VT). Myocardial fibrosis has been strongly associated with VT and sudden cardiac death (SCD) in this population [19] Additionally, a significant percentage of ACHD patients develop new atrial arrhythmias over time, emphasising its late occurrence [20]. It is of concern to note that supraventricular tachycardias may occur in up to half of ToF patients, while VT occurs in about 14% of ToF patients [21]. The prevalence of AF also appears to be steadily increasing in ACHD adults [22].

Patients with persistent left-to-right shunts such as VSD or PDA can permanently overdrive pulmonary vessels, leading to pulmonary hypertension and pulmonic stenosis, which is another quite serious complication, and in later stages, lead to Eisenmenger physiology. [23].

Beyond such hemodynamic complications, there are structural vascular issues like aortic root dilation, particularly in patients with a bicuspid aortic valve (BAV), which is also associated with Turner syndrome. This dilation progresses with age and is in part attributed to weaker aortic walls, increasing the risk of aneurysm formation or progression [24]. Often, patients with ACHD require prosthetic material or may have residual valvular pathology. These patients are susceptible to infective endocarditis (IE), which remains a persistent threat. Dental procedures have been linked to IE occurrence [25], and the overall risk of incident IE in ACHD patients is 54.8 times higher in comparison with unaffected subjects, and so, this serves as a statistic that certainly accentuates the gravity in this vulnerable group [26].

Ultimately, the physiology of ACHD is intertwined with the potential for long-term problems. Thus, understanding its trajectories and early recognition of these complex problems is very important, since prompt rhythm control, pulmonary vasodilators, endocarditis prophylaxis, and re-intervention can have major impacts on survival [26].

Echocardiography is considered the primary imaging method, valued for its convenience, immediate results, and

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capability to assess septal defects, pulmonary pressures, ventricular function, and dilation of the aortic root. Transthoracic echocardiography (TTE) is the preferred modality due to its high temporal resolution and its ability to estimate right ventricular pressures using colour Doppler [27,28] and offers better visualisation, especially in atypical anatomy. Yet, when acoustic windows are poor, cardiac magnetic resonance (CMR) becomes the gold standard, offering meticulous ventricular volume measurement, shunt (Qp: Qs) measurement and myocardial fibrosis detection. 4D-flow MRI has been shown to effectively capture and characterise multidirectional blood flow in CHD [29], further proven by a study that affirms its efficacy in Qp: Qs calculation [30]. Cardiac MRI-derived parameters, such as right ventricular end-systolic volume (RVESV) and ejection fraction (EF), show strong correlation with clinical outcomes in pulmonary hypertension, reinforcing CMR's reliability compared to invasive assessments [31].

Heart CT angiography (CCTA) is helpful when MRI is contraindicated or when vascular anatomy requires high spatial resolution, for example, coronary anomalies, aortic arch evaluation, or device planning. It is reported that CCTA outperforms invasive angiography, although [32], multidetector CT (MDCT) is a preferred noninvasive option for patients with complex surgical backgrounds or MRI contraindications [33]. When imaging is inconclusive, cardiac catheterisation remains essential for measuring pressures, assessing pulmonary vascular resistance, and defining anatomy. [34].

In the end, exercise testing, Holter/event monitoring, and laboratory evaluations (such as BNP/NT-pro BNP, liver and kidney function) are required to improve this imaging and hemodynamic framework. Furthermore, genetic screening and cooperation between a variety of specialists, including congenital cardiologists and genetic counselors, are required for comprehensive, lifelong care for ACHD [35].

V. MANAGEMENT: THERAPEUTIC INTERVENTIONS AND FOLLOW-UP RECOMMENDATIONS

- *Atrial Septal Defect:*

Repairing ASDs is not necessary until the patient exhibits signs of right ventricular dysfunction, pulmonary hypertension or arrhythmia [36]. For about 80% of patients with secundum ASDs, device closure is the preferred treatment when anatomical parameters are met (stretched diameter ≤38 mm; rim ≥5 mm, except anteriorly). Its associated with a slightly greater likelihood of reintervention, lower mortality, and a complication rate of less than 1%. While early post-procedural atrial arrhythmias are typically transient, cardiac erosion and thrombosis are rare adverse outcomes [10]. In cases where device closure is not an option, surgical closure is used to treat sinus venosus, primum, coronary sinus defect, and secundum ASDs [36]. Surgical correction of ASD has a low mortality rate and good long-term outcomes when performed early in childhood and without PH. Antiplatelet therapy (aspirin 75 mg o.d) is recommended for at least 6 months. Although it does not reduce the likelihood of arrhythmia, closure after 40 years particularly when done percutaneously improves symptoms and right cardiac function. Pre-procedural balloon occlusion and hemodynamic evaluation are essential for deciding on the appropriate closure approach (delayed, fenestrated, or complete), as ASD closure may worsen heart failure in patients with compromised left ventricular function. For individuals with PH, a comprehensive examination is required, which includes measurement of PVR (pulmonary vascular resistance). Due to the poor outcomes, closure must be avoided if PVR ≥5 WU. Fenestrated closure may be possible if L-R shunting persists and PVR falls <5 WU after PAH-targeted therapy including oral endothelin-receptor antagonist or phosphodiesterase type 5 inhibitor and subsequent hemodynamics reevaluation [37]. Additionally, it's associated with decreased PAP (pulmonary artery pressure) and improvement of symptoms. Patients with atrial arrhythmias should be evaluated for surgical ablation (e.g., modified maze) during repair. Device closure may limit future electrophysiological access to the LA, and it's essential to do a customized surgical risk assessment for elderly patients who are not candidates for device closure [10].

- *Ventricular Septal Defect:*

Surgical VSD closure is associated with good long-term outcomes and low operative risk (1-2%). Transcatheter closure is a promising alternative, especially for perimembranous, centrally situated, or residual VSDs. Risk factors observed in children, including tricuspid regurgitation, aortic regurgitation, or AV block, must be evaluated further in adults. Adult VSD closure is uncommon since the majority have minor, hemodynamically inconsequential defects or severe pulmonary hypertension, which is associated with a poor prognosis following closure [10].

- *Eisenmenger Syndrome:*

The majority of treatment centers employ a symptom-oriented, sequential approach to Eisenmenger syndrome, typically beginning with an oral ERA or PDE-5 inhibitor and increasing medication if symptoms worsen or if symptoms continue [10]. Parenteral alternatives should be actively sought if oral medication is unable to provide a sufficient improvement in symptoms. Parenteral prostaglandins work well when begun early, but subcutaneous or inhaled delivery is preferred since central intravenous catheters increase the risk of infection and paradoxical embolism in Eisenmenger patients and those with R-L shunt lesions. Following 16 weeks of treatment, endothelin receptor antagonist (bosentan) has been found to enhance 6MWT and reduce PVR in WHO functional class III Eisenmenger patients [10,38]. For patients with Eisenmenger syndrome and cyanotic patients without PAH but with an arterial oxygen saturation below 90%, pregnancy should be greatly discouraged [39]. Patients with erythrocytosis exhibiting symptoms of hyperviscosity should undergo phlebotomy [14,40].

- *Atrioventricular Septal Defect:*

Since AVSDs cannot be catheterized, surgery is required to repair the valve while closing the defect. For persistent interatrial or interventricular connections,

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endocardial pacing could be required. Patients with AVSD must be regularly monitored and assessment should include evaluation for AV valve failure, pulmonary hypertension, arrhythmias, residual shunts and ventricular enlargement. Follow-up visits should be scheduled every two to three years for patients with complete repair and minimal remaining abnormalities, depending on the severity of residual lesions [10].

- *Patent Ductus Arteriosus:*

In adults, calcification of the PDA could complicate surgical closure. Transcatheter device closure is the preferred technique since it is typically effective and has few adverse effects, even in cases where further cardiac surgery is required. In patients with aneurysms, device closure is inappropriate and they have to undergo surgery. Echocardiography should be used to evaluate residual shunt, pulmonary pressures, left ventricular size and function and associated anomalies. Patients with normal pulmonary pressures, no residual shunt and normal left ventricular function do not require routine follow-up after six months. Individuals with persistent PH or LV dysfunction should be monitored every 1-3 years depending on the condition severity [10]. Additionally, circulating vasodilatory prostaglandins may contribute to the delay or failure of closure in preterm newborns and Indomethacin is used to treat PDA in preterm newborns by suppressing the synthesis of prostaglandins [41]. Adults with PDA must have their oxygen saturation measured in both hands and feet in order to check for R-L shunting [8].

- *Coarctation of the Aorta:*

Stenting is the primary treatment, and given that covered stents have less complications, they are preferable. For children, biodegradable stents are being developed, and balloon angioplasty is utilized to dilate stented aortas again.Techniques used in pediatric surgery include bypass tube grafts, prosthetic patch aortoplasty, end-to-end anastomosis and resection. Adults with challenging anatomy may benefit from ascending-to-descending aortic conduits. Invasive evaluation is necessary for confirmation and treatment of re-coarctation when there is a greater pressure gradient (systolic ≥20 mmHg) between the upper and lower extremities.Rupture and death are possible outcomes of aneurysms at the intervention site or in the ascending aorta. Both patch repairs (eg. with Dacron) and interposition grafts should be scanned often because they are particularly susceptible to repair-site aneurysms and false aneurysms, respectively [10].

- *Bicuspid Aortic Disease:*

For asymptomatic individuals with normal aortic diameters and preserved LVEF, the watch-and-wait approach is advised. Patients with BAV have few treatment alternatives with 25% undergoing aortic replacement surgery. For bicuspid valves that are not amenable to conventional surgical techniques, TAVR might be a good substitute. Additionally, the 2020 ACC/AHA Guideline includes TAVR as a class IIb recommendation for bicuspid patients under particular circumstances. Generally, AVR (Ross procedure) is advised when the aorta diameter reaches 55 mm; however, depending on additional risk factors (Class IIa/IIb), AHA and Esc recommendations permit lower thresholds for individuals with bicuspid aortic valve (BAV)-associated aortopathy. These include the valve morphology, the degree of regurgitation, comorbidities, aortic dilatation rate, wall stress,and any family history of dissection. In certain situations, valve surgery may be advised for high-risk genetic conditions (such as Turner or Marfan) at diameters as low as 45 mm. ACEIs and beta-blockers (class IIb recommendation) are advised for BVD patients, whereas angiotensin II receptor blockers can lessen aortic root dilatation. In neonates with critical AS and reduced left ventricular systolic function, balloon valvuloplasty is usually recommended [42,43,44].

- *Tetralogy of Fallot:*

Primary neonatal ToF repair is appropriate for low-risk patients with few comorbidities and good branch pulmonary atresias (PAs). High-risk patients with small branch PAs, weights under 3 kg, and numerous comorbidities are treated with staged palliation to reduce the chance of early death. Compared to surgical palliation, catheter-based techniques are recommended. For newborns with ToF, complete surgical repair of ToF may be the first surgery performed; this usually happens between the ages of three and six months. Depending on RVOT (right ventricular outflow tract) structure and institutional preference, there are two main techniques: pulmonary valve sparing repair (VSR) and TAP repair [45]. Patients with repaired Tetralogy of Fallot (TOF) often develop pulmonary insufficiency (PI), right ventricular dilation, arrhythmias, and the risk of sudden death. Pulmonary valve replacement (PVR), using a bioprosthesis or conduit, remains the gold standard treatment for PI in these cases [46]. In recent years, transcatheter valve implantation (TPVI) using the Melody valve has emerged as a less invasive alternative to surgical PVR, especially in patients with dysfunctional RV-PA conduits or bioprosthetic valves. Branch pulmonary stenosis is a common long-term complication seen in TOF, which may need stenting [47].

- *Transposition of the Great Arteries and Fontan Circulation:*

Diuretics improve the symptoms of overt heart failure. More symptomatic people may benefit from prescriptions for "classical" heart failure medications like ARBs, ACEi, beta blockers, digoxin, even though no benefit has been shown for standard heart failure medication in patients with systemic RV failure. Given that post-capillary PH appears to be most prevalent late after atrial switch surgery, certain pulmonary vasodilator medication is contraindicated.In adults with complicated transposition (PS), VSD, and TGA, Rastelli-type repairs are frequently performed. Blood is directed from the LV to the aorta by the VSD patch, and a valved conduit connects the RV to the PA [10]. In an arterial switch operation, the reimplantation of coronary arteries from a single aortic sinus (single sinus coronary artery) is still a technically difficult technique. In this case, the coronary ostial anatomy should determine the specific coronary transfer approach [47]. Patients with atrial switch repairs may develop systemic right ventricular failure, baffle stenosis, atrial tachyarrhythmias, and sinus node dysfunction

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due to long-standing hemodynamic burden on the subaortic RV [48].

In patients with complex CHD, Fontan (in a single functional ventricle) has significantly improved life expectancy. Pacemaker implantation is frequently indicated for sinus node dysfunction or AV Block, particularly in patients with atrial switch or Fontan circulation. Whereas, in patients with ventricular arrhythmias, an epicardial or subcutaneous ICD (Implantable cardioverter-defibrillator) may be necessary [49].

- *Univentricular Heart:*

Depending on the severity of the defect, extra communications such as PDA, ASD, or VSD may be required to sustain both circulations since mixed oxygenated blood enters the systemic circulation. During the last few decades, there has been a substantial improvement in patient survival with univentricular hearts. The introduction of novel surgical palliation approaches, such as the Fontan and Norwood operations, has been followed by improvements in perioperative management and surgical refinement. When it comes to treating hypoplastic left heart syndrome (HLHS), the Norwood operation is regarded as the gold standard. The objectives of first-stage palliation are the same, despite the numerous variations in surgical technique that have been documented in the literature: unhindered systemic blood flow to the coronary arteries and aorta, a controlled source of pulmonary blood flow, and unhindered egress of pulmonary venous return. Staged palliation, branch pulmonary artery banding, the hybrid procedure, comprehensive second stage palliation, and numerous technical changes to the Norwood procedure and the superior and total cavopulmonary connections (TCPC) are just a few of the numerous technical improvements on the lengthy list [50].

- *Pregnancy:*

Pregnancy in ACHD is associated with significantly increased maternal and fetal risks, necessitating early, individualised pre-pregnancy counselling ideally initiated at a young age. Risk assessment includes ECG, echocardiography, and exercise testing, with CT or MRI as needed in complex anatomy. In high-risk cases or where pregnancy is contraindicated, early discussion should include family planning alternatives. Counselling should also include fertility, drug safety, fetal outcomes, and lifestyle factors such as smoking or alcohol use. The modified WHO classification of maternal cardiovascular risk helps in stratifying patients from Class I (no increased risk) to Class IV (pregnancy contraindicated), which helps ensure that women with high-risk such as those with fetal complications or severe ventricular dysfunction, receive appropriate guidance and multidisciplinary support in expert centres [30]. Congenital rubella is known to cause PDA, especially in regions with low immunization rates. Rubella acts as a teratogen in the early stages of pregnancy, so infection is contraindicated during pregnancy due to the risk of severe fetal anomalies [51].

- *Medical Interventions:*

Standard medical interventions that are not unique to ASD, such as antiarrhythmic medications, anticoagulation, and diuretics, are generally responsible for treating further complications, mainly arrhythmias and heart failure. For patients with atrial fibrillation (AF), established risk scores for stroke versus bleeding concerns, such as CHA2DS2-VASc and HAS-BLED, may serve as a guide for thromboprophylaxis. Direct oral anticoagulants might be taken into consideration, but there is little data supporting their usage in CHD [22,52]. There are no trials with rigorous clinical objectives specific to patients with ACHD and ACHD patients have often been excluded from adult HF clinical trials [53]. The AHA and other guidelines recommend using diuretics, beta blockers, ACE inhibitors or ARBs for ACHD with HF complications. New recommendations have been added, which now allow the use of Sodium-glucose cotransporter-2 inhibitors for the same. Similarly, the ACHD population may also benefit from the evidence supporting the use of β-blockers such as carvedilol, metoprolol, bisoprolol, and nebivolol [53].

VI. TRANSITION TO CARE

About 60% of adults with CHD experience care gaps, and just 10% obtain proper therapy [54]. It is well recognised that inconsistent care gets worse as people get older [55]. Inappropriate follow-up is frequently caused by patients who are unaware of the importance of long-term monitoring [56]. When CHD patients lose follow-up (≥3 years), it is frequently associated with worse health and higher fatality rates [57]. The American Heart Association (AHA) states that to ensure an easy transition from pediatric to adult care for CHD, transition programs must begin at age 12 [58]. Typically, teenagers with congenital heart disease require a customized transition strategy [59]. Focused instruction for this vulnerable category through structured transition programs is a crucial component of adult CHD care [60]. Chronically ill adolescents should begin transition counselling at age 12 and move into adult care by age 21 [61]. According to the AHA, to effectively self-manage cardiovascular disease, one must understand the condition and its therapy, possess the ability to use this knowledge daily and have the self-assurance to stick with these healthy habits [62].

VII. FUTURE DIRECTIONS

The strategy of care is steadily and surely maturing, as survival into adulthood becomes increasingly common among patients with ACHD. We are now seeing a shift from limited anatomical defect management to an age of advances characterised by proactive and personalised strategy. Innovations that combine imaging, artificial intelligence, regenerative medicine, and new and improved gadgets are being made possible by ongoing research. One of the most immediate and impactful changes is current technologies like three-dimensional (3D) printed heart models and virtual simulations, which are now integrated into surgical organisations. Therefore, it allows for more accurate visualisation of complex cardiac structures. These models are helpful for complex anatomy and are based on CT or MRI data. These may apply in the case of double outlet right ventricle or transposition of great arteries [31]. Similarly,

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computational modelling and holograms now play a growing role in preoperative visualisation and procedural mapping.

Beyond visualisation, Artificial intelligence (AI) is poised to revolutionise, looking forward to what could be a deterioration. Conduit stenosis and Fontan route failure are particularly worrisome. AI-based triage systems are being developed to stratify risk and guide decision-making before deterioration becomes clinically evident [63]. In concert with such innovations, gene therapy, most exceptionally the Clustered Regularly Interspaced Short Palindromic Repeats (CRISPR), now presents hope for inherited arrhythmias and channelopathies, especially long QT syndrome. Although still experimental, it shows potential. In the meantime, stem cell treatments are on the horizon. Although there is currently little clinical evidence, early uses of stem cells and cardiac progenitor cell therapy seek to enhance heart function in children with single-ventricle physiology [64]. Encouraging outcomes have also been noted with the use of mesenchymal, cardiac progenitor, and umbilical cord-derived cells (stem cells) in patients with hypoplastic left heart syndrome (HLHS) [65].

Newer and more efficient miniature devices, such as the leadless pacemaker, now provide rhythm control for ACHD patients with complicated anatomies such as interrupted venous access. There has been a triumphant implantation of the dual-chamber leadless pacemaker system in a patient with congenital heart disease, pointing to its feasibility and potential advantages over traditional pacemakers [64]. These innovations represent a major step forward in shifting ACHD management from reactive care to predictive and personalised interventions. By merging biotechnology, imaging and artificial intelligence, the new era of care holds promise for longer and healthier lives [64].

VIII. CONCLUSION

ACHD is an expanding threat that poses many challenges due to the increasing number of patients and the ever-increasing demand for advanced modalities and treatment. Identifying the type of defect, along with a detailed patient history and genetic screening, is a key feature in diagnosing ACHD. Appropriate measures to tackle future complications should be anticipated and planned accordingly to increase the quality of life of such patients. In terms of the complications and comorbidities like HF or IE, there is a significant need for proper training of health care professionals to treat and manage ACHD. Current studies have demonstrated promising results in regenerative medicine, artificial intelligence, imaging and new and better equipment. However, more research and insight into ACHD patients and their lifestyles are needed for the betterment of patient care, quick diagnosis, and early detection.

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