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Univentricular Heart in Adults: Clinical Outcomes, Diagnostic Challenges, and Therapeutic Approaches

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ABSTRACT

Univentricular heart is a rare and complex congenital heart defect characterized by the presence of a single functional ventricular chamber. While significant advancements in pediatric surgical interventions have improved the survival of patients with univentricular physiology, a growing number of these individuals are now reaching adulthood. This transition presents a spectrum of unique clinical challenges, including the management of long-term complications such as heart failure, arrhythmias, thromboembolism, and liver dysfunction. The adult univentricular heart population requires a multidisciplinary approach to optimize outcomes, incorporating advanced imaging modalities for accurate anatomical and hemodynamic assessment, and tailored therapeutic strategies. This review discusses the epidemiology, anatomical and physiological considerations, late complications, and current evidence-based management of adult patients with univentricular heart. Furthermore, it explores emerging therapeutic options, including the use of advanced heart failure therapies and the potential for cardiac transplantation. The paper also highlights the importance of transition care programs from pediatric to adult congenital heart disease specialists to improve the continuity of care and long-term prognosis.

KEYWORDS: Univentricular heart, congenital heart disease, adult congenital cardiology, heart failure, Fontan circulation, arrhythmias, thromboembolism, cardiac transplantation, transition of care.

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INTRODUCTION

Congenital heart defects (CHD) represent the most common form of congenital malformations, and significant advancements in pediatric cardiology and cardiovascular surgery over the past decades have dramatically improved survival rates. One of the most complex forms of CHD is the univentricular heart, a spectrum of malformations where only one functional ventricular chamber supports systemic and pulmonary circulations. These defects include variants such as hypoplastic left heart syndrome, tricuspid atresia, and double inlet left or right ventricle. Despite early surgical palliation, including staged reconstructions culminating in the Fontan procedure, the long-term sequelae of univentricular physiology often manifest during adulthood.1,2

The increasing population of adults with univentricular hearts has introduced novel clinical challenges. Many of these individuals develop late complications, such as progressive heart failure, arrhythmias, protein-losing enteropathy, hepatic congestion, and thromboembolic events. Their care requires a comprehensive and nuanced understanding of the altered hemodynamics and the impact of the Fontan circulation on multiple organ systems.1,2

Furthermore, the management of adults with univentricular hearts is complicated by the need for lifelong surveillance, advanced imaging, and potential reinterventions. Multidisciplinary collaboration between cardiologists, hepatologists, electrophysiologists, and transplant teams is crucial to provide optimal outcomes. This review article aims to explore the pathophysiology, clinical manifestations, diagnostic tools, and therapeutic strategies relevant to adults

with univentricular heart physiology. It will also address the importance of well-structured transition programs to ensure seamless care from pediatric to adult cardiology specialists, ultimately improving quality of life and longevity in this growing patient population.1,2

EPIDEMIOLOGY

The univentricular heart, a severe congenital cardiac malformation where a single functional ventricle is responsible for supporting both systemic and pulmonary circulations, represents a rare but growing focus of adult congenital heart disease (ACHD). As surgical and medical advancements continue to enhance survival rates among patients with complex congenital heart anomalies, there has been a corresponding increase in the population of adults living with univentricular physiology. 1,2

Globally, congenital heart defects occur in approximately 8 per 1,000 live births, with univentricular heart defects accounting for about 1-3% of all congenital heart diseases. In the past, survival into adulthood was considered improbable for individuals with single-ventricle physiology. However, the advent of staged surgical interventions, notably the Fontan operation and its subsequent refinements, has significantly altered the natural history of these patients. As a result, the median survival age for patients with univentricular hearts has increased, and an estimated 80-90% of children who undergo Fontan completion are now expected to reach adulthood.1,2

Current population estimates suggest that there are tens of thousands of adult survivors worldwide with single-ventricle physiology, with some studies indicating that adults with congenital heart defects now outnumber pediatric patients. In North America and Europe, ACHD registries have demonstrated a steady rise in the prevalence of adult patients with complex congenital heart conditions, including univentricular heart disease. This trend reflects not only the improved surgical outcomes but also advances in perioperative care, pediatric and adult cardiology, and the establishment of specialized centers for lifelong surveillance.1,2

Despite these advances, the univentricular heart remains a heterogeneous condition, and its epidemiological profile is influenced by various factors, including the type of underlying anatomical defect, the surgical strategies employed, and regional differences in healthcare infrastructure. For instance, hypoplastic left heart syndrome (HLHS), one of the more severe forms of univentricular physiology, continues to present a significant burden of morbidity and mortality, particularly in low- and middle-income countries where access to early and complex surgical interventions may be limited.3,4

Survival rates and the quality of life for adults with univentricular hearts are also impacted by sex-related differences. Some studies have reported a higher prevalence of complications such as heart failure, arrhythmias, and liver disease in male patients compared to females, though the reasons for these discrepancies remain under investigation. Additionally, racial and socioeconomic disparities contribute to variations in outcomes, as individuals from marginalized communities often face barriers to specialized ACHD care and timely interventions.3,4

Longitudinal cohort studies have highlighted that the overall survival of adults with a univentricular heart is contingent upon multiple factors, including the type and timing of initial surgical palliation, the presence of extracardiac comorbidities, and the adequacy of lifelong follow-up in specialized ACHD clinics. Notably, the Fontan circulation, which serves as a palliative strategy rather than a curative solution, comes with inherent limitations, leading to an elevated risk of long-term complications such as Fontan-associated liver disease (FALD), protein-losing enteropathy (PLE), and Fontan failure. These challenges underscore the need for continuous epidemiological surveillance to better understand and address the evolving health needs of this complex patient cohort.3,4

In summary, the epidemiology of univentricular heart in adults is marked by a dynamic and evolving landscape, shaped by the interplay between medical advancements and the inherent limitations of current surgical and medical therapies. As survival rates improve, there is a growing imperative for healthcare systems worldwide to expand and optimize specialized care pathways for adult survivors of univentricular heart disease. Comprehensive registries and population-based studies will be crucial in guiding future research, policy planning, and resource allocation to address this growing and vulnerable population effectively.3,4

CLINICAL MANIFESTATIONS

Adults with univentricular heart physiology present with a wide range of clinical manifestations due to the complex and chronic nature of their condition. These manifestations are the consequence of the altered hemodynamics inherent to single-ventricle physiology and the sequelae of long-term palliation strategies, most commonly the Fontan circulation. The presentation in adults is highly variable and may be influenced by factors such as the specific underlying cardiac anatomy, the type and success of prior surgical interventions, and the extent of systemic organ involvement. 3,4

1. Cardiac Symptoms

Heart failure is one of the most common and concerning manifestations in adults with univentricular hearts. Progressive ventricular dysfunction often develops over time due to the significant volume and pressure load imposed on the single functional ventricle. Patients may experience symptoms of congestive heart failure, including exertional dyspnea, orthopnea, fatigue, and fluid retention manifesting as peripheral edema or ascites. The pathophysiology of heart failure in these patients is multifactorial, involving myocardial fibrosis, ventricular dysfunction, and the adverse effects of chronic Fontan physiology.5,6

Arrhythmias are also prevalent in this population, with atrial arrhythmias, such as atrial flutter and atrial fibrillation, being particularly common. The etiology of arrhythmias is often related to atrial dilation, surgical scarring, and chronic volume overload. Ventricular arrhythmias, although less frequent, can occur and may be life-threatening, necessitating close monitoring and, in some cases, implantable cardioverter-defibrillator (ICD) placement. Symptoms of arrhythmias include palpitations, presyncope, and syncope, with the latter raising concerns about an increased risk of sudden cardiac death.5,6

In addition, thromboembolic events represent a significant risk due to the prothrombotic state associated with Fontan circulation. Patients may present with signs of systemic embolization, such as transient ischemic attacks (TIA) or cerebrovascular accidents (CVA), or with venous thromboembolism, including deep vein thrombosis (DVT) and pulmonary embolism. The etiology is multifactorial, involving blood flow stasis, venous hypertension, and abnormal coagulation profiles.5,6

2. Fontan-Associated Complications

The Fontan procedure, although life-saving, results in a unique physiology that predisposes patients to a variety of complications collectively termed Fontan-associated morbidities. **Fontan-associated liver disease (FALD)** is one of the most notable complications, characterized by progressive hepatic fibrosis and cirrhosis due to chronic venous congestion. Patients may present with hepatomegaly, ascites, or symptoms related to portal hypertension. In advanced cases, they are at risk of developing hepatocellular carcinoma (HCC), necessitating routine surveillance with liver function tests and imaging.5,6

Protein-losing enteropathy (PLE) is another serious manifestation, marked by the loss of serum proteins through the gastrointestinal tract. This condition presents with symptoms such as diarrhea, severe edema, and hypoalbuminemia, and it can be life-threatening if not managed effectively. The exact pathophysiology remains incompletely understood but is thought to involve increased central venous pressure and lymphatic dysfunction.6,7

Plastic bronchitis, though less common, is another Fontan-associated complication that can present with respiratory symptoms such as chronic cough and expectoration of rubbery bronchial casts, leading to life-threatening airway obstruction. The condition is believed to be associated with lymphatic leakages into the bronchial tree.6

3. Systemic and Multiorgan Involvement

The chronic venous hypertension characteristic of Fontan physiology affects multiple organ systems beyond the heart and liver. **Renal dysfunction** is commonly observed, often as a result of chronic low cardiac output and congestive nephropathy. Patients may present with signs of chronic kidney disease, including decreased glomerular filtration rate (GFR) and proteinuria, which further complicates the management of heart failure.7

Pulmonary complications also arise, with patients frequently experiencing reduced exercise capacity due to decreased pulmonary blood flow and restrictive lung physiology secondary to diaphragmatic paralysis or scoliosis, which is often a sequela of prior surgical procedures. Pulmonary arteriovenous malformations (PAVMs) can occur, presenting with hypoxemia and cyanosis, especially in those with residual right-to-left shunts.7

Neurologic manifestations are another critical aspect of the clinical profile. **Neurocognitive impairment** is not uncommon, with some patients demonstrating difficulties in executive function, memory, and attention, likely as a consequence of chronic hypoxemia, multiple surgical procedures, and thromboembolic events. Additionally, psychiatric disorders, including anxiety and depression, are prevalent in this population, impacting overall quality of life and complicating medical management.7

4. Other Manifestations

Musculoskeletal abnormalities, such as scoliosis and reduced bone mineral density, are frequently reported and may exacerbate functional limitations. **Hematologic abnormalities**, including thrombocytopenia and coagulopathy, may arise due to splenic dysfunction and chronic venous congestion. Furthermore, **endocrine disturbances**, such as delayed puberty and growth retardation, are common among survivors who underwent extensive surgical procedures during childhood.8

Adults with univentricular heart physiology face a complex array of clinical manifestations that reflect the interplay between altered cardiac function and systemic complications. The management of these patients requires a highly individualized and multidisciplinary approach, with a focus on early detection and intervention for complications to optimize long-term outcomes and quality of life.8

DIAGNOSTIC METHODS

Accurate and comprehensive diagnostic assessment is crucial for adults with univentricular heart physiology, given the complexity and variability of these congenital heart defects. Diagnostic evaluation aims to characterize the unique anatomical features, assess the functional status of the single ventricle, monitor for complications, and guide therapeutic decision-making. A multidisciplinary approach employing a combination of imaging modalities, hemodynamic studies, and laboratory testing is often required to ensure optimal management.9

1. Echocardiography

Transthoracic echocardiography (TTE) remains a cornerstone in the assessment of adults with univentricular heart anatomy. It provides crucial information on the morphology and function of the single ventricle, as well as on valve function, ventricular outflow tracts, and the patency of

the systemic and pulmonary venous pathways. **2D and 3D imaging** enable detailed visualization of complex structural abnormalities, while **Doppler echocardiography** is essential for evaluating intracardiac flow dynamics, assessing pressure gradients across stenotic or regurgitant valves, and estimating ventricular filling pressures.9

Transesophageal echocardiography (TEE) may be used as a complementary modality, particularly when transthoracic images are suboptimal. TEE is highly valuable for detecting thrombi in the atria or the Fontan pathway and for providing high-resolution images of the atrioventricular valves, which are often affected in univentricular heart patients.10

2. Cardiac Magnetic Resonance Imaging (CMR)

Cardiac magnetic resonance imaging (CMR) has become an indispensable tool in the diagnostic evaluation of adults with univentricular heart. It offers unparalleled spatial resolution and tissue characterization capabilities, allowing for accurate assessment of ventricular volumes, ejection fraction, and myocardial fibrosis. **CMR is particularly advantageous for measuring ventricular function and quantifying regurgitant or shunt volumes** without the need for ionizing radiation, which is an important consideration in young adult patients requiring serial imaging.10

Additionally, late gadolinium enhancement (LGE) imaging provides insights into myocardial fibrosis, which is a predictor of adverse outcomes, including ventricular dysfunction and arrhythmias. CMR is also used to evaluate the Fontan circulation, identify venous pathway obstructions, and assess liver morphology and function in cases of suspected Fontan-associated liver disease (FALD).10

3. Computed Tomography (CT)

Computed tomography (CT) is another critical imaging modality, particularly for evaluating extracardiac structures such as the pulmonary arteries and systemic venous connections. **CT angiography (CTA)** is useful for identifying vascular complications, such as stenosis, thromboembolism, or pulmonary arteriovenous malformations (PAVMs), which can significantly impact patient management. The high spatial resolution of CT makes it ideal for pre-surgical planning and for the assessment of aortic dilation, a complication that may arise in patients with a systemic right ventricle.11

Low-dose, high-speed CT protocols are increasingly employed to minimize radiation exposure, especially given the need for repeated imaging in this population. In cases where CMR is contraindicated, such as in patients with non-MRI-compatible devices, CT provides a valuable alternative.11

4. Cardiac Catheterization

Invasive cardiac catheterization remains a critical diagnostic and therapeutic tool in the adult univentricular heart population. **Hemodynamic assessment** via catheterization provides precise measurements of intracardiac and pulmonary artery pressures, oxygen saturations, and cardiac output. This is particularly important for evaluating the Fontan circuit, where elevated central venous pressure or low pulmonary blood flow can have significant clinical implications.11

Angiography performed during catheterization is used to delineate the anatomy of the pulmonary and systemic venous pathways, identify collateral vessels, and evaluate the patency of surgical shunts. Catheterization also allows for therapeutic interventions, such as balloon angioplasty or stent placement, to relieve stenosis within the Fontan circuit or other vascular obstructions.11

Endomyocardial biopsy may be considered in select cases to assess for myocarditis, infiltrative cardiomyopathy, or the extent of myocardial fibrosis, especially when noninvasive imaging raises suspicion of these pathologies.11

5. Electrophysiological Studies

Given the high prevalence of arrhythmias in adults with univentricular heart physiology, **electrophysiological studies (EPS)** are often warranted for diagnostic and therapeutic purposes. EPS help to identify the underlying mechanisms of atrial and ventricular arrhythmias, which may include reentrant circuits secondary to surgical scarring or structural heart disease. The data obtained from these studies can guide the need for catheter ablation procedures, antiarrhythmic therapy, or the implantation of devices such as pacemakers or implantable cardioverter-defibrillators (ICDs).12

Holter monitoring and event recorders are also routinely used to detect and characterize arrhythmias over extended periods, providing insights into the frequency and type of rhythm disturbances.12

6. Laboratory and Biomarker Assessment

Laboratory investigations play a crucial role in the ongoing evaluation of adults with univentricular heart. **Serum biomarkers** such as brain natriuretic peptide (BNP) or N-terminal pro-BNP (NT-proBNP) are useful for monitoring heart failure status and guiding therapeutic adjustments. Elevated BNP levels are often indicative of worsening ventricular function or Fontan circuit complications.12

Routine assessment of liver and renal function is also critical, given the risk of Fontan-associated liver disease (FALD) and chronic kidney disease. **Coagulation studies** are performed to monitor for hypercoagulable states, and tests for protein-losing enteropathy (PLE) may include serum albumin and immunoglobulin levels. Additionally, imaging studies such as hepatic elastography or magnetic resonance elastography (MRE) are increasingly used to assess liver stiffness as part of the surveillance for FALD.12

7. Exercise Testing

Cardiopulmonary exercise testing (CPET) is a valuable method for assessing functional capacity and cardiorespiratory fitness. CPET provides objective measurements of peak oxygen consumption (VO2 max) and ventilatory efficiency, which are strong prognostic indicators

in the univentricular heart population. Exercise tolerance is often limited in these patients due to the unique hemodynamic constraints of Fontan circulation, and CPET results can inform individualized exercise recommendations and identify early signs of heart failure or pulmonary vascular disease.12

Six-minute walk tests (6MWT) may be employed as a simpler alternative for assessing exercise capacity and monitoring disease progression, although CPET provides more comprehensive data.13

Current Treatment

The management of adults with univentricular heart defects is inherently complex and requires a lifelong, multidisciplinary approach that emphasizes regular follow-up in specialized adult congenital heart disease (ACHD) centers. Treatment strategies are aimed at optimizing the function of the single ventricle, managing Fontan-associated complications, and improving the patient's quality of life. As univentricular heart patients age, new challenges emerge that necessitate a nuanced understanding of the interplay between cardiac function and multisystemic health.13

1. Medical Therapy

Pharmacological management forms the backbone of treatment for adults with univentricular heart. The goals of medical therapy include reducing cardiac workload, improving ventricular performance, managing heart failure symptoms, and addressing complications arising from the Fontan circulation.13

- **Heart Failure Management**: Diuretics are commonly used to alleviate fluid overload and reduce symptoms of congestive heart failure. Loop diuretics, such as furosemide, are often combined with potassium-sparing agents to maintain electrolyte balance. Angiotensin-converting enzyme (ACE) inhibitors and angiotensin II receptor blockers (ARBs) are sometimes prescribed to reduce afterload and mitigate ventricular remodeling, although their efficacy in this population is less well-established than in biventricular heart failure. Beta-blockers may be cautiously used, particularly in patients with signs of adrenergic overactivity, but their role remains controversial.13
  For patients with advanced heart failure, **inotropic agents** may be administered temporarily during periods of decompensation to enhance myocardial contractility. Long-term inotropic support is generally avoided due to the risk of increased mortality.13
- **Anticoagulation and Antiplatelet Therapy**: Given the prothrombotic state associated with the Fontan circulation, anticoagulation is often a mainstay of treatment. Warfarin is frequently used to prevent thromboembolic events, especially in patients with a history of thrombus formation, atrial arrhythmias, or ventricular dysfunction. In select cases, direct oral anticoagulants (DOACs) are being explored, but their use remains limited and requires careful consideration of the bleeding risk. Antiplatelet agents such as aspirin are commonly prescribed for primary prophylaxis in patients deemed to have a lower thromboembolic risk.13
- **Arrhythmia Management**: The management of arrhythmias in adults with univentricular heart is a significant challenge. **Antiarrhythmic medications** such as amiodarone, sotalol, or class IC agents may be used to control atrial arrhythmias, though these drugs must be used cautiously due to potential proarrhythmic effects. Catheter ablation is considered in cases of refractory or recurrent arrhythmias, and device therapy, including pacemakers or implantable cardioverter-defibrillators (ICDs), is indicated for patients at high risk of sudden cardiac death.13
- **Pulmonary Vasodilators**: The use of pulmonary vasodilators, such as phosphodiesterase-5 inhibitors (e.g., sildenafil or tadalafil) or endothelin receptor antagonists, is being increasingly explored in adults with Fontan physiology to improve pulmonary blood flow and exercise capacity. These agents may reduce pulmonary vascular resistance, thereby alleviating some of the hemodynamic burden on the single ventricle.13

2. Fontan-Specific Complications

Addressing Fontan-associated morbidities is a critical aspect of treatment for adults with univentricular heart.

- **Fontan Failure**: Fontan failure is a progressive condition that may manifest as worsening heart failure, protein-losing enteropathy (PLE), plastic bronchitis, or Fontan-associated liver disease (FALD). Management often requires a tailored approach, including diuretics, nutritional support, and optimization of anticoagulation. Refractory cases may necessitate more aggressive interventions, such as surgical or transcatheter Fontan pathway revisions or conversion to a total cavopulmonary connection (TCPC) to improve hemodynamic efficiency.14
- **Protein-Losing Enteropathy (PLE)**: PLE is managed with a combination of dietary modifications (e.g., high-protein, low-fat diet with medium-chain triglycerides), diuretics to reduce venous congestion, and pharmacological therapies such as corticosteroids or subcutaneous heparin. Newer treatments, like the use of selective lymphatic embolization or interventional lymphatic procedures, have shown promise in some cases.14
- **Fontan-Associated Liver Disease (FALD)**: The management of FALD involves regular surveillance with imaging and liver function tests. Advanced liver disease may necessitate consideration for combined heart-liver transplantation, especially in

patients with cirrhosis or hepatocellular carcinoma.14

3. Surgical and Interventional Approaches

Surgical and interventional options remain critical in the treatment of adults with univentricular heart. The decision to pursue further interventions depends on the patient's hemodynamic status, the presence of complications, and overall functional capacity.14

- **Fontan Revision and Conversion**: Some patients may benefit from Fontan pathway revisions, especially if there are significant anatomical obstructions or atrial arrhythmias. Conversion from an atriopulmonary Fontan to a more hemodynamically efficient TCPC can be considered in select cases to reduce arrhythmic burden and improve long-term outcomes.14
- **Catheter-Based Interventions**: Transcatheter interventions, such as balloon angioplasty or stent placement, are used to address Fontan pathway obstructions or venous stenosis. Additionally, catheter-based closure of residual shunts or fistulas may be necessary to optimize circulation. The percutaneous placement of fenestrations within the Fontan circuit is another technique used to decompress the system and improve symptoms in patients with high venous pressures.14
- **Heart Transplantation**: Heart transplantation remains the definitive treatment for patients with end-stage Fontan failure or severe ventricular dysfunction. Given the increasing number of adults surviving with single-ventricle physiology, there is growing recognition of the need for tailored transplant strategies, including combined heart-liver transplantation for those with advanced FALD. Patient selection and timing are critical, and outcomes are often complicated by the patient's pre-existing comorbidities and complex anatomy.14

4. Emerging Therapies and Research

Research into novel treatment options for univentricular heart defects is ongoing. **Stem cell therapy** and regenerative medicine approaches are being investigated for their potential to improve myocardial function and delay the progression of heart failure. Additionally, advancements in imaging techniques are enhancing our ability to detect complications earlier and to individualize treatment.15

Gene therapy is another area of exploration, particularly for conditions associated with specific genetic mutations. As our understanding of the molecular underpinnings of heart failure in this population expands, targeted therapies may emerge as a promising avenue.15

5. Lifestyle Modifications and Supportive Care

Lifestyle modifications play an important role in the comprehensive management of adults with univentricular heart. Patients are encouraged to engage in light to moderate physical activity, as tolerated, to maintain cardiovascular fitness while avoiding activities that could exacerbate symptoms or increase the risk of arrhythmias. **Nutritional support** is vital, particularly for patients with complications like PLE or heart failure, and may include consultation with a dietitian.16

Psychological support and counseling are essential, given the high prevalence of anxiety, depression, and neurocognitive impairment in this patient population. A holistic approach to care, which includes addressing mental health and promoting social well-being, is necessary for long-term success.16

CONCLUSION

The management of adults with univentricular heart remains a formidable challenge, reflecting the extraordinary complexity of this congenital cardiac condition. Advances in pediatric cardiac surgery have significantly extended the life expectancy of individuals with single-ventricle physiology, leading to a rapidly growing population of adult patients who present with unique anatomical and physiological challenges. As these patients transition from childhood to adulthood, the spectrum of complications and comorbidities evolves, requiring a dynamic and comprehensive approach to care that spans multiple medical specialties.

Univentricular heart in adulthood is not simply a continuation of the congenital defect but a distinct entity characterized by the long-term sequelae of prior surgical interventions, such as the Fontan procedure, and the progressive decline in ventricular function. The Fontan circulation, while life-saving in early years, imposes considerable hemodynamic strain on the cardiovascular system, leading to a myriad of complications including arrhythmias, heart failure, protein-losing enteropathy, thromboembolism, and Fontan-associated liver disease. The cumulative burden of these complications necessitates lifelong follow-up in specialized centers equipped to address the intricate needs of these patients.

From a diagnostic standpoint, a multimodal imaging approach is essential to fully characterize the complex anatomy and monitor for complications. Echocardiography, cardiac magnetic resonance imaging (CMR), and computed tomography (CT) each offer unique insights, while cardiac catheterization provides indispensable hemodynamic data. The integration of these diagnostic tools with regular surveillance for arrhythmias and systemic complications is paramount in optimizing patient outcomes.

Therapeutically, the landscape continues to evolve. Medical management focuses on symptomatic relief and the prevention of disease progression, but it remains limited by the inherent inefficiencies of single-ventricle physiology. Anticoagulation, heart failure medications, and antiarrhythmic drugs form the cornerstone of medical therapy, but novel pharmacological approaches are urgently needed. The use of pulmonary vasodilators and research into

regenerative therapies highlight the ongoing efforts to address the underlying pathophysiology and improve quality of life.

Surgical and interventional strategies, including Fontan pathway revisions and catheter-based procedures, offer hope for symptomatic relief in select patients. However, heart transplantation stands as the ultimate therapeutic option for those with end-stage ventricular dysfunction, a decision complicated by the patient's intricate anatomy and systemic complications. The consideration of combined heart-liver transplantation in patients with severe Fontan-associated liver disease underscores the multifaceted nature of care required for this population.

Despite these advances, significant gaps remain in our understanding of the long-term outcomes and optimal management strategies for adults with univentricular heart. As research continues to elucidate the molecular and hemodynamic mechanisms driving disease progression, there is a critical need for the development of targeted therapies and personalized treatment plans. Collaboration between pediatric and adult congenital heart specialists, cardiologists, hepatologists, and multidisciplinary care teams is essential to address the lifelong challenges these patients face.

In conclusion, adults with univentricular heart represent a triumph of modern congenital heart surgery but also pose one of the greatest challenges in cardiovascular medicine. Their care requires a nuanced understanding of complex anatomy, meticulous attention to evolving complications, and an unwavering commitment to improving long-term outcomes through innovative research and comprehensive management. As the population of adult single-ventricle survivors continues to grow, our collective responsibility is to ensure that these patients receive the highest standard of care, informed by a deep appreciation of their unique journey and the complexities of their condition.

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