Review

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Review

Subjectivity and Surveillance Gaps in Post-Fontan Assessment and Their Impact on Late Heart Transplant Referral

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Abstract

The Fontan procedure represents the final stage of palliation for patients with single-ventricle congenital heart disease and has enabled survival into adulthood for a growing population of patients. However, the Fontan circulation is inherently non-physiological, characterized by chronically elevated systemic venous pressure and reduced cardiac output, which predispose patients to progressive circulatory dysfunction and multiorgan complications. Despite increasing recognition of Fontan circulatory failure, referral for heart transplantation frequently occurs late in the disease course, often after irreversible end-organ damage has developed. Several factors contribute to this delay. Surveillance of ventricular function is largely dependent on echocardiography, which is frequently limited by anatomical complexity and inter-observer variability, leading to subjective assessment of ventricular performance. In addition, inconsistent use of invasive hemodynamic assessment and variable implementation of multimodality imaging may delay detection of early circulatory deterioration. Equally important, extracardiac complications—including Fontan-associated liver disease and renal dysfunction—are often under-recognized or inadequately monitored, despite their major impact on transplant candidacy and outcomes. This review examines the limitations of current surveillance strategies in Fontan patients and highlights how subjectivity in cardiac assessment and gaps in systemic monitoring may contribute to delayed transplant referral. A more structured, multidisciplinary surveillance approach incorporating objective imaging and systematic end-organ assessment may facilitate earlier recognition of Fontan failure and improve timing of transplant evaluation.

Keywords: Fontan surveillance; subjectivity; FALD; Fontan associated nephropathy

1. Background

The Fontan procedure represents the final stage of palliation for patients with complex congenital heart disease characterized by a single functional ventricle. Although the operation has dramatically improved survival into adolescence and adulthood, it does not constitute a curative repair. Instead, the Fontan circulation establishes a unique physiological state characterized by chronically elevated systemic venous pressure and reduced cardiac output. Over time, this non-physiological hemodynamic configuration predisposes patients to progressive circulatory inefficiency, ventricular dysfunction, and a spectrum of multi-organ complications affecting the liver, kidneys, lymphatic system, and other organ systems [1].

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Despite major advances in surgical techniques and perioperative care, long-term outcomes remain limited. In the current era, approximately 5%–10% of Fontan patients will die or require heart transplantation during childhood, and nearly 50% are expected to face death or transplantation by the age of 40 years. These statistics highlight the progressive nature of Fontan physiology and underscore the importance of timely recognition of circulatory failure and appropriate referral for advanced therapies [2].

Late referrals of patients with single-ventricle physiology for heart transplantation remains a major challenge in contemporary pediatric and adult congenital cardiology. Several studies suggest that a substantial proportion of Fontan patients are referred for transplant evaluation only after significant clinical deterioration has already occurred. In some cohorts, up to 40% of Fontan patients are referred late in the course of disease, and a notable proportion may be declined for transplantation shortly after referral due to advanced end-organ dysfunction. Such delays reflect the difficulty in recognizing early Fontan failure and emphasize the need for objective and structured surveillance strategies capable of identifying deterioration before irreversible systemic complications develop [3,4].

The trajectory toward Fontan circulatory failure is highly variable, and the timing of referral for transplant evaluation can significantly influence listing decisions and outcomes. However, current transplantation guidelines provide limited objective criteria to guide the optimal timing of referral in this unique population. Furthermore, the characteristics and outcomes of Fontan patients who are evaluated but ultimately not listed for transplantation remain poorly described in the literature, leaving an important knowledge gap regarding their prognosis and clinical management. This article aims to explore the factors contributing to delayed referral of Fontan patients for heart transplantation and to discuss the current framework for systematic surveillance that may help bridge this gap. By highlighting the limitations of existing assessment strategies and emphasizing the need for more objective markers of Fontan deterioration, we seek to contribute to a more proactive approach to the identification and management of patients approaching Fontan failure [5–7].

Several systemic challenges contribute to delayed recognition of advanced Fontan physiology. First, the transition of care from pediatric cardiology to adult congenital heart disease services remains a vulnerable period during which continuity of surveillance may be disrupted and subtle signs of circulatory deterioration may be overlooked. Second, the multisystem nature of Fontan disease requires clinicians to monitor extracardiac complications—particularly hepatic and renal dysfunction—yet many cardiologists have limited experience in the interpretation and longitudinal monitoring of these organ-specific assessments. Finally, uncertainty regarding the optimal diagnostic strategies for surveillance, including the appropriate use of cardiac imaging modalities, cardiopulmonary testing, and structured evaluation of liver and renal function, often leads to variability in follow-up practices and delayed identification of clinically significant complications. Addressing these challenges is essential to improving the timely recognition of Fontan failure and facilitating earlier referral for transplant evaluation[8].

2. Main Body

2.1. Types of Heart Failure in Fontan Patients

The long-term burden of the Fontan circulation is largely driven by its fundamentally abnormal hemodynamic design. Unlike the normal cardiovascular system, in which a sub-pulmonary ventricle actively pumps blood through the pulmonary circulation, the Fontan circuit relies on passive flow of systemic venous blood into the pulmonary arteries. Pulmonary blood flow therefore depends on a pressure gradient in which central venous pressure must exceed pulmonary arterial pressure. Although this arrangement permits pulmonary perfusion in the absence of a right ventricle, it simultaneously imposes chronic systemic venous hypertension while limiting ventricular preload and effective cardiac output. This inherent hemodynamic contradiction—requiring elevated venous

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pressure to maintain pulmonary flow while simultaneously impairing forward circulation—is often referred to as the **"Fontan paradox."** [9].

Over time, the Fontan circulation operates near the limits of physiological compensation. Even small increases in pulmonary vascular resistance, ventricular diastolic stiffness, or atrioventricular valve regurgitation can significantly impair forward flow. As a result, patients frequently develop progressive circulatory inefficiency characterized by reduced cardiac output during both rest and exercise, impaired ventricular filling, and systemic venous congestion. This delicate balance contributes to the gradual development of what is commonly termed **Fontan circulatory failure** [1,10].

Importantly, Fontan patients are at risk for two distinct yet overlapping forms of heart failure. The first resembles conventional ventricular systolic dysfunction, in which deterioration of myocardial contractility leads to reduced ejection performance and symptomatic heart failure. The second reflects **failure of the Fontan circulation itself**, which may occur even in the presence of relatively preserved ventricular systolic function. In this scenario, elevated systemic venous pressure, impaired ventricular preload, lymphatic congestion, and reduced systemic perfusion collectively drive clinical deterioration. This form of circulatory failure is increasingly recognized as a hallmark of late Fontan physiology and is often difficult to detect using traditional measures of ventricular function alone [11].

The pathophysiological consequences of chronic Fontan hemodynamics extend far beyond the heart. Progressive venous congestion and impaired lymphatic drainage contribute to a wide spectrum of extracardiac complications, including **protein-losing enteropathy, plastic bronchitis, hepatic fibrosis and cirrhosis, renal dysfunction, and systemic inflammatory activation**. These systemic manifestations not only worsen quality of life but also complicate the management of advanced disease and significantly influence candidacy for heart transplantation [12].

Among the cardiovascular complications, **arrhythmias represent one of the most frequent and clinically significant problems in the post-Fontan population**. Both sinus node dysfunction and atrial tachyarrhythmias occur commonly, particularly in patients with atrio-pulmonary or lateral tunnel Fontan modifications. Progressive atrial dilation, surgical scarring, and abnormal hemodynamics create a highly arrhythmogenic substrate. As a consequence, a substantial proportion of patients ultimately require permanent pacemaker implantation or catheter ablation procedures during long-term follow-up [13].

Importantly, the development of arrhythmias is not merely an isolated electrical complication but often serves as a marker of deteriorating Fontan physiology. Persistent tachyarrhythmias can exacerbate circulatory inefficiency by impairing ventricular filling and reducing effective cardiac output, while bradyarrhythmias related to sinus node dysfunction may further limit exercise capacity and systemic perfusion. Numerous studies have demonstrated that the presence of recurrent arrhythmias is strongly associated with worsening ventricular function, increased hospitalizations, and a higher risk of transplantation or death. Consequently, arrhythmias often represent a pivotal turning point in the clinical trajectory of Fontan patients and may herald the transition toward advanced circulatory failure [14].

Understanding the complex interplay between abnormal hemodynamics, ventricular dysfunction, and systemic complications is therefore essential for recognizing the early stages of Fontan deterioration. As the burden of end-stage Fontan physiology continues to grow in the expanding adult congenital heart disease population, timely identification of patients progressing toward advanced heart failure remains a critical challenge and a key determinant of transplant eligibility and long-term outcomes.

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2.2. Subjectivity of Echocardiographic Assessment in Assessment Of Single Ventricle Failure

Echocardiographic Surveillance and Its Limitations

Echocardiography remains the most widely used imaging modality for routine surveillance in patients with Fontan circulation. Current clinical guidelines recommend annual echocardiographic evaluation as part of long-term follow-up in order to monitor ventricular performance, atrioventricular valve function, and the integrity of the Fontan pathway. The widespread availability, portability, and absence of radiation exposure make echocardiography an attractive first-line tool for longitudinal assessment in this complex population. However, despite its central role in clinical practice, echocardiographic assessment in Fontan patients is inherently limited and frequently subject to substantial variability [15].

One of the principal challenges arises from the **anatomical and geometric complexity of single-ventricle physiology**. Standard methods used to quantify ventricular volumes and ejection fraction are based on geometric assumptions derived from normal biventricular anatomy. In patients with single-ventricle circulation, the dominant ventricle may be morphologically left- or right-sided, frequently with irregular shapes and altered contraction patterns following multiple surgical interventions. As a result, traditional volumetric measurements often lack accuracy and reproducibility when applied to these morphologically abnormal ventricles [16].

In addition to these geometric limitations, several technical factors can further compromise image quality. Many Fontan patients undergo multiple thoracic surgeries during infancy and childhood, resulting in surgical scarring, altered thoracic anatomy, and chest wall deformities that may impair acoustic windows. As patients age, increasing body habitus, pulmonary hyperinflation, and skeletal changes can further degrade image quality, particularly in adolescents and adults. Consequently, complete visualization of the ventricular cavity and Fontan pathway may be difficult to achieve during routine transthoracic examinations [17].

These technical and anatomical challenges contribute to **substantial inter-observer variability in the assessment of ventricular function**. Numerous studies have demonstrated only modest agreement between observers when visually estimating ventricular performance in morphologic left ventricles and even weaker agreement in morphologic right ventricles. Because quantitative measurements are often unreliable or technically difficult to obtain, clinicians frequently rely on qualitative visual estimation—commonly referred to as "eyeballing"—to judge ventricular contractility. While this approach may provide a rapid clinical impression, it introduces a considerable degree of subjectivity into clinical decision-making and may contribute to delayed recognition of progressive circulatory deterioration [15,18].

Advanced echocardiographic techniques, including **speckle-tracking strain imaging**, have been proposed as potential tools to reduce subjectivity and improve reproducibility in the assessment of myocardial function. Global longitudinal strain and other deformation indices may detect subtle myocardial dysfunction before changes in ejection fraction become apparent. However, the application of these techniques in Fontan patients is not without limitations. Differences between imaging platforms, vendor-specific software algorithms, and variability in tracking quality can lead to inconsistent results across institutions. Furthermore, strain analysis in morphologically abnormal ventricles remains technically challenging and may not always be feasible in patients with poor acoustic windows. Consequently, although advanced echocardiographic methods provide valuable additional information, they do not fully eliminate the subjective component of ventricular assessment in the Fontan population [19,20].

2.3. Role of Multimodality Imaging

Given the limitations of echocardiography, contemporary surveillance strategies increasingly emphasize the importance of **multimodality imaging** in the evaluation of Fontan patients. Combining complementary imaging techniques allows clinicians to obtain a more comprehensive understanding of ventricular function, vascular anatomy, and hemodynamic performance[21].

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Cardiac magnetic resonance imaging (CMR) is widely considered the **reference standard for the assessment of ventricular volumes and function** in Fontan circulation. CMR provides highly accurate and reproducible measurements of ventricular size, mass, and ejection fraction without relying on geometric assumptions. In addition, phase-contrast imaging enables detailed quantification of blood flow through the Fontan pathway, pulmonary arteries, and systemic veins. CMR can also evaluate collateral vessels, detect thrombus formation, and assess myocardial fibrosis through techniques such as late gadolinium enhancement and T1 mapping. These capabilities make CMR an invaluable tool for comprehensive evaluation of Fontan physiology [22].

Despite these advantages, CMR is not universally feasible for all patients. Younger children may require general anesthesia or deep sedation to tolerate the prolonged imaging protocol. Arrhythmias, irregular breathing patterns, and implanted devices can compromise image quality. Additionally, some patients with metallic implants or severe claustrophobia may be unable to undergo the examination. Access to specialized CMR expertise may also be limited in certain centers. For these reasons, echocardiography continues to serve as the primary surveillance modality in many clinical settings, with CMR often performed at periodic intervals rather than annually [22,23].

Cardiac computed tomography (CT) may serve as an alternative imaging modality when CMR is contraindicated or unavailable. Modern CT scanners provide high-resolution anatomical visualization and are particularly useful for evaluating the Fontan pathway, pulmonary arteries, systemic venous structures, and implanted stents or conduits. CT imaging can also help delineate extracardiac anatomy and detect collateral vessels that may contribute to cyanosis or volume overload. However, its role in routine surveillance remains limited due to concerns regarding radiation exposure, the need for iodinated contrast administration, and the relatively limited ability of CT to assess ventricular function compared with CMR [24,25].

2.4. Inconsistent Use of Cardiac Catheterization

Another area of significant variability in the follow-up of Fontan patients is the use of **diagnostic cardiac catheterization**. Although non-invasive imaging modalities provide valuable structural and functional information, cardiac catheterization remains the gold standard for direct measurement of hemodynamic parameters, including central venous pressure, pulmonary artery pressure, ventricular end-diastolic pressure, and pulmonary vascular resistance. These measurements are critical for understanding the physiological performance of the Fontan circulation and for identifying potentially reversible contributors to circulatory failure. Despite its diagnostic value, there is currently **no universal consensus regarding the optimal frequency of routine invasive hemodynamic assessment** in Fontan patients. Some surveillance programs advocate scheduled catheterization at defined intervals—often approximately every ten years—or prior to major transitions in care, such as the transition to adult congenital heart disease services. Others recommend catheterization only when clinical symptoms or non-invasive imaging findings suggest deterioration [21].

Common indications for catheterization include worsening cyanosis, progressive exercise intolerance, refractory arrhythmias, unexplained ventricular dysfunction, or evidence of hepatic congestion. In addition to diagnostic evaluation, catheterization may provide opportunities for therapeutic interventions, such as angioplasty or stent placement for pathway obstruction, embolization of collateral vessels, or fenestration creation in selected cases. However, in many clinical settings catheterization is performed **only when overt clinical deterioration becomes apparent**. As a result, subtle hemodynamic changes—such as gradually rising Fontan pressures or increasing pulmonary vascular resistance—may remain undetected until the disease has progressed significantly. This variability in surveillance strategies contributes to delays in identifying early Fontan failure and may ultimately influence the timing of referral for advanced therapies, including heart transplantation [24–26].

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2.5. Systemic Congestion and Complications

The systemic consequences of Fontan physiology are increasingly recognized as major determinants of long-term morbidity and mortality. However, two important observations continue to shape clinical practice in ways that may delay recognition of advanced disease. First, many cardiologists tend to focus primarily on **cardiac function and the classical complication of protein-losing enteropathy**, often overlooking the broader spectrum of extracardiac manifestations associated with chronic venous congestion. As a result, complications affecting other organ systems—particularly the liver, kidneys—may remain under-recognized until they reach advanced stages[27]. Second, the evaluation of these systemic complications frequently falls outside the traditional expertise of cardiologists, leading to variability in surveillance strategies and delays in appropriate diagnostic testing. These factors contribute to the underestimation of the true systemic burden of Fontan physiology and may ultimately influence the timing of referral for advanced therapies such as heart transplantation [28].

2.6. Fontan Associated Liver Disease (FALD)

Fontan-associated liver disease has emerged as one of the most important long-term complications. Progressive hepatic fibrosis may develop silently and can eventually progress to cirrhosis and hepatocellular carcinoma [29].

Surveillance of Fontan-associated liver disease (FALD) requires a multimodal approach, as no single test reliably detects early disease or accurately reflects fibrosis severity. Among biochemical markers, **gamma-glutamyl transferase (GGT)** is often the earliest abnormal laboratory parameter, reflecting early cholestasis and hepatic congestion, although its lack of specificity limits its diagnostic value when used in isolation. Other liver enzymes such as AST and ALT may become mildly elevated but correlate poorly with the degree of fibrosis, while markers of hepatic synthetic function—including bilirubin, albumin, and INR—typically become abnormal only in advanced disease30. Consequently, imaging plays a central role in surveillance. **Abdominal ultrasound** remains the most widely used first-line imaging modality due to its availability and ability to detect parenchymal heterogeneity, hepatomegaly, and signs of portal hypertension. **Ultrasound elastography** can provide additional information by measuring liver stiffness and may detect abnormalities early, but its interpretation is complicated by the confounding effects of hepatic congestion inherent to Fontan physiology. **Magnetic resonance elastography (MRE)** offers more reproducible assessment of liver stiffness and may better characterize fibrosis, although its cost and limited availability restrict routine use31. Cross-sectional imaging with **CT or MRI** is particularly valuable for detecting advanced structural changes such as cirrhosis or hepatic nodules, but these modalities generally identify later manifestations of disease. While **liver biopsy** remains the histological gold standard for staging fibrosis, its invasive nature and potential sampling error limit its use as a routine surveillance tool [32].

Importantly, the complexity and limitations of these diagnostic tools may contribute to the **under-recognition of progressive hepatic disease in Fontan patients**, particularly in clinical settings where surveillance protocols are not standardized. Many cardiologists may rely primarily on routine cardiac imaging while liver assessment is performed inconsistently or interpreted outside a structured framework. As a result, hepatic dysfunction may progress silently for years before becoming clinically apparent, at which point advanced fibrosis, cirrhosis, or portal hypertension may already be present. This delayed recognition of systemic complications may ultimately influence **the timing of referral for heart transplantation**, as patients may present for transplant evaluation only after significant and sometimes irreversible end-organ damage has developed [33–35].

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Table 1. Comparison of different markers of FALD [29–31].

Modality / Test	Role in Surveillance	Earliest Abnormal Finding	Strengths	Limitations
Serum GGT	**Earliest biochemical marker of FALD**	Elevation may appear early due to chronic hepatic congestion and cholestasis	Inexpensive, widely available, easily repeatable, useful for longitudinal monitoring	Nonspecific; can be elevated due to medications or biliary disease; does not quantify fibrosis or structural liver damage
Other liver enzymes (AST, ALT)	Routine biochemical surveillance	Often mildly elevated later than GGT	Widely available and easy to monitor	Poor correlation with fibrosis severity; may remain normal despite advanced FALD
Bilirubin / albumin / INR	Indicators of hepatic synthetic function	Usually **late findings**	Reflect advanced liver dysfunction	Insensitive for early disease; abnormal values often indicate advanced pathology
Ultrasound abdomen	**First-line imaging screening test**	Can detect early parenchymal heterogeneity or hepatomegaly	Widely available, non-invasive, inexpensive, can identify nodules and portal hypertension	Operator dependent; limited sensitivity for early fibrosis
Ultrasound elastography (FibroScan / SWE / ARFI)	Early detection of increased liver stiffness	Elevated stiffness may occur early after Fontan	Non-invasive fibrosis assessment; useful for trend monitoring	Liver stiffness influenced by congestion; cannot reliably distinguish congestion from fibrosis; no validated cutoffs
Magnetic resonance elastography (MRE)	Advanced fibrosis assessment	Potentially earlier detection of fibrosis than standard imaging	More accurate than ultrasound elastography in many settings; reproducible	Expensive, limited availability; still affected by congestion
MRI or CT liver imaging	Structural assessment	Detects cirrhosis, nodules, and HCC	Excellent anatomical detail; MRI preferred for nodule characterization	Detects relatively late structural changes; CT involves radiation
Liver biopsy	Gold standard for fibrosis assessment	Detects histological fibrosis	Provides definitive staging of fibrosis	Invasive; sampling error; not practical for routine surveillance

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2.7. Renal Dysfunction in Fontan, superiority of Cystatin over Creatinine

Renal dysfunction represents another **frequently under-recognized systemic complication of the Fontan circulation**. The abnormal hemodynamic environment created by the Fontan physiology—characterized by chronically elevated central venous pressure and reduced cardiac output—has profound effects on renal perfusion and function. Chronic venous congestion increases renal venous pressure, while the reduction in cardiac output limits effective renal arterial perfusion. The resulting decrease in renal perfusion pressure, defined as the difference between mean arterial pressure and central venous pressure, leads to progressive impairment in glomerular filtration and contributes to the development of chronic kidney disease in this population [36].

In addition to these hemodynamic disturbances, neurohormonal activation plays an important role in Fontan-associated renal dysfunction. Reduced renal perfusion activates the renin–angiotensin–aldosterone system, sympathetic nervous system, and vasopressin pathways, leading to sodium retention, fluid accumulation, and further venous congestion. These mechanisms create a **self-perpetuating cycle of congestion and renal injury**, similar to the cardiorenal syndrome observed in advanced heart failure. Over time, these pathophysiological mechanisms contribute to progressive deterioration in renal function [37].

Recent longitudinal data suggest that renal dysfunction is both **early and progressive in patients with Fontan circulation**. In a cohort study of Fontan patients followed for more than a decade, renal function—measured by creatinine-based estimated glomerular filtration rate (eGFR)—declined at an average rate of approximately **1.36 mL/min/1.73 m² per year**, a rate faster than that observed in healthy individuals. Importantly, deterioration in renal function was associated with increased mortality, highlighting the clinical significance of renal impairment as a marker of Fontan circulatory failure [38].

However, the early detection of renal dysfunction in this population remains challenging. Traditional markers such as **serum creatinine frequently underestimate the degree of renal impairment**, particularly in Fontan patients who often have reduced skeletal muscle mass and smaller body habitus. As a result, creatinine-based estimations of glomerular filtration rate may overestimate true renal function and delay recognition of kidney injury. Biomarkers such as **cystatin C**, which are less influenced by muscle mass, may provide a more accurate estimate of renal function and have been shown to identify a higher prevalence of renal dysfunction compared with creatinine-based measurements [39].

Despite these insights, systematic renal surveillance is not consistently incorporated into many Fontan follow-up programs. Renal dysfunction may therefore progress silently for years before being clinically recognized. Increasing evidence suggests that progressive decline in renal function may serve as an **important marker of systemic Fontan failure**, reflecting the broader multiorgan consequences of the abnormal Fontan hemodynamics. Early identification of renal impairment may therefore provide an opportunity for earlier intervention and may also inform the timing of referral for advanced therapies such as heart transplantation [40].

2.8. The Current Consensus in Fontan Surveillance

The current clinical practice algorithm for follow-up of patients with Fontan circulation emphasizes structured longitudinal surveillance beginning immediately after the procedure and continuing throughout childhood, adolescence, and adulthood. In the first year following Fontan completion, patients undergo frequent clinical evaluations every three to six months including physical examination, oxygen saturation measurement, electrocardiography, and transthoracic echocardiography to monitor early postoperative complications such as pleural effusions, ventricular dysfunction, atrioventricular valve regurgitation, arrhythmias, and cyanosis. In addition to routine clinical monitoring, preventive strategies are recommended for all patients, including antiplatelet or anticoagulation therapy, lifestyle counseling, and education regarding potential Fontan-related complications [41].

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Beyond the first postoperative year, surveillance expands to include **multisystem evaluation**, reflecting the progressive nature of Fontan physiology. During childhood and adolescence, follow-up visits are typically performed every 6–12 months with ECG, echocardiography, and oxygen saturation monitoring. Additional periodic assessments include ambulatory rhythm monitoring, cardiopulmonary exercise testing, and laboratory testing such as BNP/NT-proBNP levels, comprehensive metabolic panels, GGT, cystatin C, and hematologic studies. Imaging surveillance is also recommended, including cardiac magnetic resonance imaging every two to three years when feasible and abdominal imaging with ultrasound and elastography to detect early Fontan-associated liver disease. Diagnostic catheterization is generally reserved for clinical deterioration or performed at longer intervals, typically beginning approximately ten years after Fontan completion [42].

In adulthood, the surveillance algorithm becomes increasingly comprehensive, reflecting the rising burden of systemic complications and the growing risk of Fontan failure. Follow-up every 6–12 months includes cardiac imaging, biomarker monitoring, and evaluation of liver and renal function. Multimodality imaging—such as cardiac MRI, abdominal MRI with elastography, cardiopulmonary exercise testing, and rhythm monitoring—is recommended at regular intervals to assess ventricular performance, Fontan pathway anatomy, and extracardiac complications. The algorithm also highlights the importance of multidisciplinary care, including hepatology referral, pulmonary function testing, and consultation with heart failure or transplant specialists when appropriate. These recommendations underscore the recognition that Fontan circulation is a multisystem condition requiring coordinated long-term surveillance rather than isolated cardiac monitoring [43].

3. Conclusions

The long-term management of patients with Fontan circulation remains complex. The reliance on subjective echocardiographic assessment, combined with inconsistent use of invasive hemodynamic evaluation and variable surveillance of liver and kidney complications, contributes to delayed recognition of Fontan failure. These gaps in monitoring can ultimately lead to late referral for heart transplantation. A more structured and objective surveillance strategy—supported by multimodality imaging, systematic end-organ monitoring, and multidisciplinary collaboration—may improve the timely identification of patients who would benefit from transplant evaluation. Such an approach is essential to optimize long-term outcomes for the growing population of Fontan survivors.

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Abbreviations:

Abbreviation	Full Term
ALT	Alanine Aminotransferase
ARFI	Acoustic Radiation Force Impulse
AST	Aspartate Aminotransferase
BNP	B-type Natriuretic Peptide
CMR	Cardiac Magnetic Resonance Imaging
CPET	Cardiopulmonary Exercise Testing
CT	Computed Tomography
eGFR	Estimated Glomerular Filtration Rate
FALD	Fontan-Associated Liver Disease
GGT	Gamma-Glutamyl Transferase
HCC	Hepatocellular Carcinoma
INR	International Normalized Ratio
MRI	Magnetic Resonance Imaging
MRE	Magnetic Resonance Elastography
NT-proBNP	N-terminal pro-B-type Natriuretic Peptide
SWE	Shear Wave Elastography

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