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# Transcatheter Edge-to-Edge Repair in Adult Congenital Heart Disease: Anatomic Challenges, Device Adaptations, and Emerging Evidence

REVIEW

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## ABSTRACT

Atrioventricular valvular regurgitation (AVVR) is a principal determinant of morbidity and mortality in adults with congenital heart disease (ACHD), and surgical management carries substantial operative risk owing to prior sternotomies, distorted intracardiac anatomy, impaired ventricular function, and lesion-specific hemodynamic fragility. Transcatheter edge-to-edge repair (TEER)—using MitraClip or TriClip (Abbott) or PASCAL/PASCAL ACE (Edwards Lifesciences)—has emerged as a compelling alternative in high-surgical-risk acquired valvular disease and is rapidly expanding into ACHD. We review the anatomic substrate, device-specific technical considerations, preprocedural imaging requirements, and clinical evidence supporting TEER across the principal ACHD substrates: congenitally corrected transposition of the great arteries (TGA) with systemic tricuspid regurgitation, atrial switch repairs for dextro-TGA, atrioventricular septal defects, and single-ventricle Fontan circulations. We also address procedural challenges unique to ACHD, including venous access anomalies, altered transseptal geometry, absent conventional echocardiographic windows, and iatrogenic stenosis risk in morphologically abnormal valves. Multidisciplinary selection integrating ACHD structural expertise, advanced imaging, and procedural TEER experience is requisite for safe outcomes.

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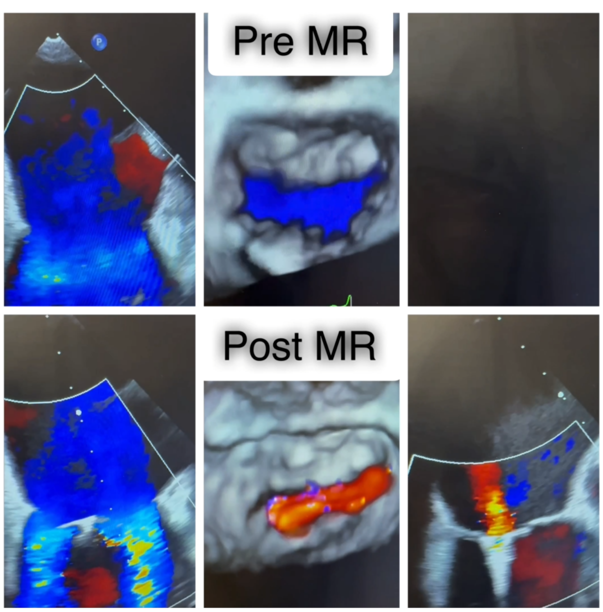
## INTRODUCTION

More than one million adults in the United States are living with adult congenital heart disease (ACHD) since decades of improving surgical palliation have transformed once lethal lesions into chronic conditions requiring lifelong surveillance.[1,2] Atrioventricular valvular regurgitation (AVVR) is among the most consequential late sequelae, contributing disproportionately to heart failure hospitalizations, ventricular remodeling, arrhythmias, and premature mortality.[3] Congenital AV valve complexity including leaflet dysplasia, abnormal chordal architecture, inlet ventricular septal defects (VSDs), and aberrant annular geometry renders surgical repair technically demanding, with elevated operative risk in patients with prior surgery, limited reserve, and residual lesions.[4]

Transcatheter edge-to-edge repair (TEER) was developed for percutaneous treatment of mitral regurgitation (MR) in acquired disease based on the Alfieri stitch concept of creating a double-orifice configuration via leaflet coaptation.[5] Following EVEREST II (Endovascular Valve Edge-to-Edge Repair Study) and the landmark COAPT (Cardiovascular Outcomes Assessment of the MitraClip Percutaneous Therapy for Heart Failure Patients with Functional Mitral Regurgitation) trials, Abbott's MitraClip-based TEER is now well established for primary and secondary MR in high-surgical-risk patients.[6,7] The TRILUMINATE (Trial to Evaluate Cardiovascular Outcomes in Patients Treated with the Tricuspid Valve Repair System) Pivotal study led to the Food and Drug Administration (FDA) approval of TriClip (Abbott) for tricuspid TEER (T-TEER) in April 2024.[8] PASCAL (Edwards Lifesciences), approved for both mitral and tricuspid applications, offers an alternative clip-and-spacer design.[9]

Case reports and single-center series now document TEER across diverse ACHD substrates, including congenitally corrected transposition of the great arteries (ccTGA), dextro-TGA (d-TGA) after atrial switch, Fontan palliation, and complete atrioventricular canal (CAVC) defect with encouraging procedural success and short-term outcomes that nonetheless demand careful interpretation given the heterogeneity and rarity of these lesions.[10,11] This review synthesizes existing evidence, lesion-specific anatomic challenges, preprocedural planning requirements, and TEER's role within a multidisciplinary ACHD structural heart program (Video 1).




**Video 1** Echocardiograms, including 3D TEEs, of atrioventricular valve before and after TEER developed for percutaneous treatment of mitral regurgitation; also see at https://vimeo.com/1200546282. MR: mitral regurgitation; TEEs: transesophageal echocardiograms; TEER: transcatheter edge-to-edge repair

## AV VALVULAR REGURGITATION PATHOPHYSIOLOGY IN ACHD

Mechanisms of AVVR in ACHD are distinct from acquired disease and vary across lesions. Three predominate: (1) annular dilation from chronic volume overload or atrial enlargement; (2) intrinsic leaflet and subvalvular dysplasia, such as with Ebstein anomaly and complete atrioventricular septal defects (AVSD); and (3) ventricular geometric distortion impairing papillary muscle alignment with leaflet tethering, analogous to functional MR in ischemic cardiomyopathy but amplified by discordant ventriculoarterial connections and non-standard trabecular anatomy.[12]

In ccTGA, the morphologic tricuspid valve serves as the systemic AV valve and is ill-suited for systemic pressures: It progressively dilates as the mRV enlarges under systemic afterload, with Ebstein-like dysplasia in 20% to 30% of cases.[4,13] TR is a major driver of mRV failure with adverse prognosis when severe. In d-TGA after atrial switch (Mustard/Senning), the subpulmonary LV may develop progressive dysfunction with associated morphologic mitral regurgitation, particularly with elevated pulmonary vascular resistance or ventricular-ventricular interaction.

In CAVC defects, the common AV valve or its post-repair components regurgitate through cleft leaflet malcoaptation, annular dilation, leaflet retraction, and suture dehiscence.[3] The Fontan circulation is uniquely intolerant of AVVR: elevated Fontan pressures (typically 12-18 mm Hg), absent pulmonary pulsatility, and low ventricular preload narrow the compensation window, and even moderate AVVR can precipitate protein-losing enteropathy, plastic bronchitis, or progressive Fontan failure.[14]

## TEER DEVICE PLATFORMS: DESIGN PRINCIPLES RELEVANT TO ACHD

### MitraClip/TriClip

Abbott's MitraClip system (fourth generation, G4) consists of a 24F steerable guide, a clip delivery system, and cobalt-chromium clips with polyester fabric. G4 offers four clip sizes (NT, NTW, XT, XTW) with variable arm length and width, enabling tailored treatment across leaflet anatomies. Independent leaflet grasping, introduced in G4, permits selective capture and repositioning particularly relevant in ACHD, where leaflet asymmetry and abnormal coaptation are common.[15] TriClip (Abbott) is anatomically homologous but configured for tricuspid anatomy, with a modified delivery catheter allowing greater anterior angulation and torquability for right-sided access.

## PASCAL/PASCAL ACE

PASCAL (Edwards Lifesciences) employs a central spacer flanked by independently articulating paddles and clasps; the spacer fills the coaptation gap directly, reducing leaflet tensioning forces and theoretically reducing leaflet trauma and iatrogenic mitral stenosis. PASCAL ACE, a smaller-profile version, suits valves with restricted leaflet mobility or limited coaptation length. In the CLASP IID (Edwards PASCAL Transcatheter Valve Repair System Pivotal) clinical trial, PASCAL was non-inferior to MitraClip on combined safety and efficacy in primary MR at prohibitive surgical risk.[9] Independent leaflet grasping without simultaneous bilateral capture is conceptually advantageous in ACHD, where dysplasia limits uniform leaflet presentation.

## PREPROCEDURAL PLANNING AND MULTIMODALITY IMAGING

Preprocedural evaluation is substantially more complex than for acquired disease and requires multimodality imaging, hemodynamic assessment, and simulation. A heart team, ACHD cardiologists, structural interventionalists, ACHD surgeons, and imagers with congenital expertise are essential.[10]

### TRANSTHORACIC AND TRANSESOPHAGEAL ECHOCARDIOGRAPHY

Two- and three-dimensional (3D) transesophageal echocardiography (TEE) is the cornerstone of intraprocedural guidance. Standard windows and nomenclature for acquired mitral and tricuspid anatomy are often inapplicable in ACHD. In ccTGA, the systemic tricuspid valve sits leftward and posterior to the normal mitral position with apical septal-leaflet displacement (Ebstein-like) and frequently three or more leaflets with variable commissural anatomy.[13] In CAVC, segmental analysis of the bridging leaflets defines the regurgitant jet and target coaptation zone. A standardized ACHD-adapted protocol with 3D reconstruction and commissural mapping should be performed at a center with congenital echo expertise.

Quantification of regurgitation severity must account for the absence of validated grading criteria for morphologically atypical valves. 3D color Doppler vena contracta area is more robust than conventional jet area or proximal isovelocity surface area, particularly for the eccentric, noncircular regurgitant orifices common in ACHD.[11]

### CARDIAC MAGNETIC RESONANCE IMAGING

Cardiac magnetic resonance (CMR) provides complementary and often indispensable data, including ventricular volumetric quantification, regurgitant fraction measurement, and tissue characterization. In Fontan patients, CMR-derived single ventricle end-diastolic volume index, ejection fraction, and AVVR by flow mapping guide procedural risk stratification. CMR-derived 3D datasets can generate patient-specific anatomic models for simulation, validated at several pediatric and ACHD centers to facilitate clip trajectory planning for novel valve geometries.[16]

### CARDIAC COMPUTED TOMOGRAPHY

Cardiac CT angiography (CCTA) characterizes vascular access, venous anomalies, and intracardiac spatial relationships. In post-surgical patients, the inferior vena cava (IVC) may be interrupted (left isomerism/heterotaxy), lateralized, or obstructed by baffles, conduits, or patch material, affecting catheter deliverability and transseptal approach. CCTA also delineates atrial baffle anatomy in post-Mustard/Senning patients, informing whether standard femoral venous access to the systemic venous atrium is feasible and whether intra-baffle puncture is achievable.[17]

### HEMODYNAMIC ASSESSMENT

Comprehensive hemodynamic profiling is requisite. Fontan pressures, transpulmonary gradient, pulmonary vascular resistance, ventricular end-diastolic pressures, and systemic venous hypertension inform candidacy and risk. With Fontan pressures > 18 to 20 mm Hg or severely depressed systemic ventricular function, decompensation risk is high and TEER should be approached cautiously or as a transplant bridge.[14]

## LESION-SPECIFIC APPLICATIONS OF TEER IN ACHD

### CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

The systemic morphologic TV in ccTGA is the most commonly reported ACHD substrate for TEER, with the largest published case experience.[10,11] It features a leftward, superior septal leaflet insertion with Ebstein-like displacement in a significant minority, a subvalvular apparatus comparable to acquired functional TR, and a globular, nonplanar annulus. The primary TR mechanism is annular dilation from mRV dilatation under systemic afterload, often compounded by leaflet tethering. (Table 1)

TriClip is most often used, deployed at the anterior-septal coaptation gap, the dominant regurgitant zone in most reported cases. Right femoral venous access and transseptal puncture into the morphologic left atrium permit clip delivery into the systemic tricuspid valve. This geometry requires careful atrial delineation and may necessitate customized guide-catheter shaping or a deflectable sheath.[10]

A five-center series in the January 2026 *JACC: Advances* reported on 20 ACHD patients undergoing TEER, of whom 60% had systemic tricuspid valves, demonstrated 95% reduction of AVVR to mild-to-moderate (1+ to 2+) at 30 days, with NYHA III–IV rates falling from 90% at baseline to 10.5%.[11] One procedure-related death (5%) from intractable heart failure occurred within 30 days. These data, although limited by sample size, support TEER feasibility in ccTGA and as a bridge to transplantation or palliative stabilization in inoperable patients.[10,13]

### D-TGA AFTER ATRIAL SWITCH: SYSTEMIC TRICUSPID REGURGITATION AND TRANSBAFFLE TEER

Among the most surgically complex ACHD substrates for TEER is d-TGA palliated by the Mustard or Senning atrial switch (predominantly 1970s-1980s), which redirected systemic and pulmonary venous return via intra-atrial baffles to restore a physiologically, albeit not anatomically corrected, circulation. The mRV thus supports the systemic circulation indefinitely, and progressive mRV dysfunction with systemic TR are the cardinal late complications driving mortality in this aging cohort.[18] In the ACHD registry of 1,168 adults followed a median of 9.2 years, moderate-to-severe TR was present in 24% at baseline and 64% among those meeting the composite end point of death, transplantation, or mechanical circulatory support, with both severe systemic RV dysfunction and moderate-severe TR independently associated with adverse outcomes on multivariable analysis.[19] A separate cohort study classified patients by risk at age 30 and demonstrated transplant-free survival of only 31% by age 45 in the “high-risk” group (defined by moderate-severe SRV dysfunction or TR) versus 87% in the low-risk group, underscoring the prognostic weight of TR in this population.[20]

Surgical correction carries prohibitive risk in most adults due to reoperation through calcified pericardium, impaired SRV function, and the difficulty of operating on a valve under systemic pressure in a reoperative field with prior baffle material. Tricuspid valve replacement, the usual procedure, requires cardiopulmonary bypass with baffle protection and carries in-hospital mortality of 5% to 15% with moderate or greater SRV dysfunction.[21] This surgical risk profile has generated considerable interest in percutaneous TEER as an alternative or bridge strategy, particularly given the established transbaffle access precedent from electrophysiologic catheter ablation in this population.[22]

The defining technical challenge of systemic TV TEER after atrial switch is transbaffle puncture to access the morphologic right-sided atrium (now the neopulmonary venous atrium) contiguous with the systemic mRV. From right femoral venous access, catheters traverse the systemic venous baffle (which carries IVC and superior vena cava blood to the mLV) and must cross the baffle material into the pulmonary venous atrium to reach the morphologic TV. The baffle is a surgically constructed partition of GORE-TEX, Dacron, or native pericardium; Mustard repairs (more common in North America) are typically more rigid and less deformable than Senning repairs, which use native atrial tissue. Puncture site selection requires meticulous preprocedural imaging: CCTA defines baffle thickness, calcification, proximity to the pulmonary venous atrium, and spatial relationships to the right pulmonary veins, tricuspid annulus, and coronary sinus.[17]

Iriart et al. reported a landmark case of systemic TV TEER via transbaffle approach after Mustard repair.[23] A 48-year-old with severely impaired SRV function underwent preprocedural 3D TEE and CCTA; under fluoroscopic and 3D TEE guidance, the baffle was punctured with a standard transseptal needle, and a single MitraClip XTR achieved

| SUBSTRATE/TARGET VALVE | MECHANISM OF REGURGITATION | ACCESS & IMAGING | DEVICE & TECHNIQUE |
|---|---|---|---|
| ccTGA — systemic TR<br>Systemic morphologic tricuspid valve (mTV) | Annular dilation - mRV dilation under systemic afterload<br>Ebstein-like dysplasia in 20-30% | Transseptal into morphologic LA<br>Trajectory orthogonal to usual mitral axis<br>3D TEE with commissural mapping | Wide-arm clips at anteroseptal gap |
| d-TGA, atrial switch — systemic TR (transbaffle TEER)<br>Systemic mTV and mRV | Progressive systemic mRV failure with TR<br>Moderate to severe TR ~24% to ~64% at death/ transplant/MCS | Transbaffle puncture<br>CCTA + 3D ICE<br>Close access with septal occluder | Mirror-image valve orientation<br>Check baffle pressures + gradient |
| d-TGA, atrial switch — subpulmonary MR<br>Subpulmonary mMV | Functional MR from subpulmonary LV dysfunction<br>Elevated PVR/PA pressure | Via pulmonary venous baffle limb<br>No conventional transseptal needed<br>Assess fixed vs post-capillary PVR | Target antero-rightward mMV<br>Standard clip platforms |
| AVSD/CAVC<br>Left "mitral" component of repaired common AV valve | Cleft malcoaptation, annular dilation<br>Patch/suture dehiscence | Standard transseptal to left-sided valve<br>Segmental 3D echo + simulation | Wide-arm clips for dilated annulus<br>Multiple clips across cleft; preserve orifice |
| Single-ventricle Fontan<br>Systemic AV valve(s) of single ventricle | AVVR poorly tolerated in passive circulation<br>Drives PLE, plastic bronchitis, Fontan failure | Conduit puncture or fenestration dilation<br>CMR + full hemodynamics<br>Caution if Fontan pressure > 18–20 mm Hg | Often multiple clips<br>Avoid ↑PVR/↓preload; sparing pacing |

**Table 1 Lesion-specific considerations for transcatheter edge-to-edge repair (TEER) in adult congenital heart disease.** AVVR: atrioventricular valvular regurgitation; AVSD/CAVC: atrioventricular septal defect/complete atrioventricular canal; CCTA: cardiac CT angiography; ccTGA: congenitally corrected transposition; CMR: cardiac MR; d-TGA: dextro-transposition; ICE: intracardiac echocardiography; LA: left atrium; MCS: mechanical circulatory support; mMV/mRV/mTV: morphologic mitral/right-ventricular/tricuspid valve; MR/TR: mitral/tricuspid regurgitation; PA: pulmonary artery; PLE: protein-losing enteropathy; PVR: pulmonary vascular resistance; RF: radiofrequency; TEE: transesophageal echocardiography

significant TR reduction. The access was closed with an 8 mm ASD occluder to prevent paradoxical embolism. Sustained NYHA improvement and SRV volume reductions were documented at 1 and 6 months.[23]

The transbaffle puncture itself requires adaptations beyond the standard transseptal technique. The baffle material does not exhibit the same tactile and echocardiographic "tent" deformation as the native fossa ovalis, often benefiting from radiofrequency-assisted puncture systems (such as the VersaCross RF wire system (Boston Scientific) or Baylis radiofrequency transseptal needle (Baylis Medical Technologies) to safely traverse the baffle. Precise fluoroscopic targeting, using biplane angulation with simultaneous right anterior oblique and left anterior oblique projections, coregistered with CCTA overlay, is essential to confirm perpendicular baffle entry without posterior wall perforation or coronary sinus injury. 3D intracardiac echocardiography (ICE) is increasingly preferred over TEE in this substrate, as ICE catheters within the systemic venous atrium provide unobstructed visualization of the baffle and adjacent structures without the geometric limitations of TEE probe orientation in d-TGA anatomy.[10]

After transbaffle access, the MitraClip/TriClip guide is steered into the morphologic right-sided pulmonary venous atrium, directed superiorly and rightward toward the systemic morphologic TV. Valve orientation relative to the delivery system is the mirror image of standard configuration; the morphologic TV faces leftward and inferiorly, opposite the standard mitral TEER trajectory. Operators must mentally reorient fluoroscopic and echocardiographic views and communicate using congenital valve nomenclature (anterior, posterior, septal leaflets of the morphologic TV) rather than acquired tricuspid segmental language. Wide-arm clip configurations (XTW or NTW) are preferred for the typically dilated morphologic TV annulus. Post-clip assessment should include the transmitral gradient across the morphologic TV (to exclude iatrogenic stenosis) and systemic venous baffle pressures (to detect significant residual left-to-right shunting that could diminish systemic ventricular preload). Routine closure of the transbaffle access site with a septal occluder is often required, as residual baffle defects carry risk of systemic thromboembolism and paradoxical embolism in a population already prone to atrial arrhythmias and ventricular dysfunction.

## D-TGA AFTER ATRIAL SWITCH: SUBPULMONARY LEFT VENTRICULAR DYSFUNCTION AND MITRAL REGURGITATION

The subpulmonary mLV and its associated mitral valve in d-TGA after atrial switch occupy a physiologically distinct position: they support the pulmonary circulation and are connected to the pulmonary venous atrium via the mMV, with the mLV ejecting into the pulmonary artery at subsystemic pressures. In contrast to the systemic mRV, which bears the full burden of systemic afterload, the subpulmonary mLV is chronically pressure-unloaded and subject to volume-related remodeling. Mitral regurgitation in this context, arising from the mMV, is encountered in a subset of patients, most commonly in the context of progressive subpulmonary LV dysfunction with secondary (functional) annular dilation, elevation in pulmonary vascular resistance causing elevated pulmonary artery pressures with consequent increased afterload on the mLV, or concomitant intrinsic mMV pathology. When severe, subpulmonary MR exacerbates pulmonary venous hypertension, worsens right-sided hemodynamics via ventricular-ventricular interaction through the shared interventricular septum, and can accelerate the progression of systemic mRV failure, creating a vicious cycle of biventricular dysfunction.

Subpulmonary mMV TEER requires an entirely different access strategy from systemic morphologic TV TEER. The pulmonary venous atrium, which receives oxygenated pulmonary venous return and is contiguous with the mLV, is accessed via the pulmonary venous baffle limb from a standard femoral venous approach; no conventional transseptal puncture is required because access is obtained directly through the venous baffle that channels systemic venous blood into the mLV. The delivery system navigates to the mMV, which lies in a rightward, anterior position, the anatomic inverse of its typical orientation in acquired mitral disease. When transseptal puncture is required for access to the mitral position valve, it traverses the baffle partition between systemic and pulmonary venous atria with target trajectory toward the anterior-rightward mMV rather than the left sided mTV.[17]

Preprocedural assessment must evaluate the pulmonary vascular bed; if pulmonary vascular resistance is fixed rather than post-capillary, mLV unloading may paradoxically unmask RV-PA uncoupling and worsen systemic mRV output. With a dominant post-capillary component, MR reduction should alleviate pulmonary venous hypertension, reduce mLV volume burden, and improve systemic mRV geometry via interventricular interaction, analogous to right heart responses to left-sided MR reduction in acquired functional MR.[24] Careful preprocedural hemodynamic provocation testing, including vasodilator challenge and temporary MR simulation via rapid pacing to reduce regurgitant fraction, can help discriminate reactive from fixed pulmonary hypertension and stratify procedural risk in this uniquely complex substrate. The published ACHD TEER literature to date does not include dedicated case series of subpulmonary mMV TEER in d-TGA, underscoring the rarity of this indication.

## ATRIOVENTRICULAR SEPTAL DEFECTS

AVVR in CAVC defects, both the repaired and unrepaired forms, result from a fundamentally different valve architecture, which is a five-leaflet common atrioventricular valve (superior and inferior bridging leaflets, left and right mural leaflets, and an anterosuperior leaflet) that spans both ventricles. After surgical repair, residual or recurrent regurgitation typically originates from cleft mitral leaflet malcoaptation, annular dilation, or patch dehiscence. TEER in this context targets the left-sided "mitral" component of the repaired common valve, positioning clips across the cleft or at the zone of maximal coaptation failure.[16]

The absence of normal mitral coaptation geometry with shortened leaflets, an inlet VSD (in unrepaired form), and shallow subvalvular space creates challenges for adequate leaflet capture. Simulation and preprocedural 3D echocardiographic reconstruction are particularly valuable to confirm anatomic eligibility. Iatrogenic mitral stenosis risk is heightened given the restricted baseline mitral valve orifice area in many repaired CAVC patients; a transmitral mean gradient > 5 mm Hg post-clip should prompt immediate reassessment of clip configuration.

## SINGLE-VENTRICLE FONTAN CIRCULATIONS

Fontan TEER is the frontier of ACHD structural intervention. The passive Fontan circulation means any procedural complication or worsening AVVR can precipitate hemodynamic crisis, yet severe AVVR drives protein-losing enteropathy, plastic bronchitis, and accelerated Fontan failure, and surgery carries operative mortality of 10% to 15% in the reoperative setting.[14]

Access for TEER in Fontan patients varies by Fontan type. In extracardiac Fontan conduits, the native atrium is accessible via a direct conduit puncture approach or, if a fenestration is present, via dilation of the existing fenestration to accommodate the 24F guide catheter. The first reported case of mitral TEER in an adult extracardiac Fontan patient used this fenestration-dilation strategy successfully.[25] In lateral tunnel Fontans, the native atrium lies within the tunnel and permits more conventional catheter delivery. The Jolley series (Boston Children's) first standardized simulation-guided

TEER protocol in CHD, including Fontan, and achieved successful MitraClip placement in all four subjects, with sustained AVVR reduction and symptomatic improvement at 30 days.[16]

Anesthesia must avoid agents that raise pulmonary vascular resistance or reduce preload. Adenosine or rapid ventricular pacing routine in standard TEER should be used sparingly given Fontan intolerance of transient cardiac-output reductions. A second venous access line in the Fontan circuit permits continuous pressure monitoring.

## PROCEDURAL CONSIDERATIONS AND TECHNICAL ADAPTATIONS

### VASCULAR ACCESS AND TRANSSEPTAL PUNCTURE

Standard TEER uses right femoral venous access with transseptal catheterization under TEE and fluoroscopic guidance targeting the posteroinferior fossa ovalis. In ACHD, this paradigm is frequently modified: venous anomalies (persistent left SVC, bilateral SVCs, interrupted IVC with azygos continuation, occluded venous segments) may necessitate left femoral, transhepatic, internal jugular, or alternative access.[3]

The transseptal puncture site must be individually tailored to the target valve geometry. For systemic TV TEER in ccTGA, the puncture is directed toward the floor of the morphologic left atrium at a position that provides adequate catheter reach to the leftward systemic AV valve, a trajectory orthogonal to the conventional mitral TEER axis. In post-Mustard/Senning repairs, a puncture through the interatrial baffle is required and demands real-time 3D guidance to avoid posterior baffle perforation. Prior residual or patch-closed atrial septal defects may serve as alternative access points. ICE guidance as an adjunct or replacement for TEE in cases of limited acoustic windows—particularly in patients with situs inversus, dextrocardia, or extensive prior thoracic surgery or hardware from valves, leads, and ventricular assist devices—should be part of the procedural toolkit at experienced ACHD TEER centers.

### CLIP SELECTION AND DEPLOYMENT STRATEGY

Wide-arm MitraClip configurations (XTW, NTW) are preferred for large coaptation gaps or dilated annuli, common in ccTGA and AVSD; PASCAL ACE's spacer may better fill gaps in valves with restricted leaflet mobility. Independent leaflet grasping should be maximized for symmetrical capture, and a post-clip mean gradient > 5 mm Hg across a mitral-position valve should prompt withdrawal.

Multiple clips are often required for large coaptation gaps in Fontan or elongated cleft defects in CAVC, with sequential deployment preserving the central orifice area to avoid functional stenosis. Commissural deployment (anteroseptal/anterolateral) in tricuspid-position TEER can address eccentric jets, although single-leaflet detachment risk is higher at commissural positions.

### INTRAPROCEDURAL HEMODYNAMIC MONITORING AND END POINTS

Post-clip assessment includes transmitral/transtricuspid gradients, downstream filling pressures, and Fontan circuit pressure when applicable. Color Doppler targets MR/TR ≤ 2+, although in ACHD with reduced ventricular compliance, reduction to moderate regurgitation may still confer meaningful benefit. Success is defined by ≥ 1-grade AVVR reduction without procedural death, embolization, emergent surgery, or new severe iatrogenic stenosis.

## CURRENT EVIDENCE BASE

The evidence supporting TEER in ACHD remains largely observational, confined to case reports, case series, and small multicenter retrospective cohorts. No randomized controlled trial has been performed or is currently underway in this population. Use in ACHD should be approached within the framework of an institutional review process, with informed consent emphasizing the investigational nature of the intervention.

In the most comprehensive multicenter experience to date, Perl et al. included 20 ACHD patients across five centers (median age 49.3 years),[11] with all targeting the systemic circulation (60% tricuspid). Procedural success (AVVR ≤ 2+ at 30 days) was 95%, with significant NYHA improvement at 30 days and 6 months; 30-day mortality was 5%, from intractable heart failure rather than procedural complication. These align with prior reports, including Alshawabkeh et al., the first ACHD mitral TEER case series; Guerin et al., the first descriptive cohort for systemic AVVR in CHD; and Jolley et al., a standardized simulation-guided pediatric TEER protocol with favorable outcomes.[10,16,26]

TRILUMINATE Pivotal (win ratio 1.48 for TriClip versus medical therapy, sustained quality-of-life benefit at 1 year) provides a framework for the hemodynamic benefits of TR reduction.[8] However, ACHD differs fundamentally from TRILUMINATE subjects (with mean age 78, predominantly functional TR, non-congenital morphology), and direct extrapolation is unwarranted without dedicated registry data.

## PATIENT SELECTION FRAMEWORK

Absent randomized data, selection requires individualized consideration by the heart team. According to general eligibility criteria from published experience and expert consensus, anatomic eligibility should be confirmed by 3D TEE and supplemented with CMR or CCTA. Minimum leaflet length for clip capture is generally ≥ 7 to 10 mm, for coaptation depth is ≤ 11 mm for mitral-position valves, and the absence of severe leaflet dysplasia or calcification precluding reliable grasping. A valve area that permits safe clip placement without generating a mean gradient > 5 mm Hg post-clip must be anticipated.

**Clinical criteria:** Symptomatic severe AVVR (NYHA II–IV) or asymptomatic severe AVVR with ventricular dysfunction, progressive dilation, or AVVR-attributable Fontan pressure elevation; high or prohibitive surgical risk; no coexisting lesion requiring non-deferrable surgery; no active endocarditis or significant leaflet destruction.

**Hemodynamic contraindications:** Fontan pressures > 20 mm Hg without AVVR as a dominant contributor; severely impaired systemic ventricular function (EF < 20%) with likely fixed low output; severe pulmonary arterial hypertension in biventricular patients when right-heart failure is pressure-driven rather than volume-driven.

## COMPLICATIONS AND RISK MITIGATION

Complications mirror acquired-disease TEER but face greater hemodynamic fragility. Single-leaflet detachment (SLD), the most common failure mode, may precipitate acute decompensation in patients with limited reserve; immediate recognition by 3D TEE with clip repositioning or withdrawal is appropriate. SLD incidence is 2% to 4% in experienced hands in acquired disease and may be higher in ACHD given atypical leaflet morphology.

Iatrogenic mitral stenosis is a particular concern in AVSD with small baseline valve areas; serial monitoring of post-clip gradients is required, and a resting mean gradient > 5 mm Hg with symptoms of elevated LA pressure may require clip retrieval.

Transseptal access complications, including pericardial tamponade, baffle perforation in atrial switch patients, and conduit injury in Fontan patients, underscore the need for meticulous procedural planning. Cardiac surgical backup must be available in the interventional suite, and detailed written protocols for emergent access to the pericardial space and cardiopulmonary bypass initiation should be established before each ACHD TEER procedure.

## FUTURE DIRECTIONS

Device miniaturization, particularly guide catheters below 24F, would broaden access in small-body patients and facilitate navigation through restrictive fenestrations or baffles. Artificial intelligence-enhanced intraoperative imaging and patient-specific 3D-printed simulators integrated into standard protocols may reduce failure rates in complex anatomy.[16]

A dedicated ACHD TEER registry with congenital anatomic classification is urgently needed to generate natural history, a benchmark, and outcome data informing selection criteria and device development. The Adult Congenital Heart Association, American College of Cardiology Congenital Heart Disease Committee, and Structural Heart Disease Council have expressed interest in formalizing access pathways and developing training standards for operators combining ACHD and structural interventional expertise.

Longer-term durability data beyond 6 to 12 months are essential to characterize clip fatigue, leaflet response, and ventricular remodeling trajectory. Whether sustained AVVR reduction translates into the COAPT-observed survival benefit, or whether ACHD physiology modifies this relationship, remains a critical open question.

## CONCLUSION

Transcatheter edge-to-edge repair represents a paradigm-shifting therapeutic option for atrioventricular valvular regurgitation in adult congenital heart disease, a population historically underserved by transcatheter structural interventions and at high risk for surgical reintervention. The existing evidence, though limited by sample size and observational design, consistently demonstrates the technical feasibility of TEER across diverse ACHD substrates, including ccTGA, Fontan circulations, atrial switch repairs, and atrioventricular septal defects, with meaningful short-term reductions in regurgitation severity and symptomatic burden. Procedural success demands a rigorous, multidisciplinary approach, comprehensive multimodal preprocedural planning, anatomically adapted delivery techniques, and center-level experience in both structural TEER and ACHD. The field now requires prospective registry infrastructure, standardized anatomic eligibility criteria, operator training pathways, and longer-term outcome data to fulfill the promise suggested by these early results.

## KEY POINTS

- Atrioventricular valve regurgitation is a major driver of morbidity and mortality in adults with congenital heart disease (ACHD), and traditional surgical repair is often high-risk due to prior surgeries, complex anatomy, and reduced ventricular function.
- Transcatheter edge-to-edge repair, using devices like MitraClip, TriClip, and PASCAL, has expanded from acquired valve disease into ACHD, offering a less invasive option with encouraging procedural success and symptom improvement in high-risk patients.
- ACHD patients require detailed preprocedural evaluation (transesophageal echocardiography, cardiac magnetic resonance, computed tomography) and specialized heart teams due to unique challenges such as abnormal valve structures, altered cardiac geometry, and difficult vascular access.
- Early case series and small cohorts show high procedural success and symptomatic benefit, but evidence remains observational; long-term outcomes, standardized selection criteria, and dedicated registries are still needed.

## COMPETING INTERESTS

The author has no competing interests to declare.

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