SASCI-MAYO CLINIC FELLOWS WEBINAR

SASCI-Mayo Clinic Fellows webinar: Lifelong management of adults with repaired tetralogy of Fallot

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ABSTRACT

This is the second in our series of South African Society of Cardiovascular Intervention (SASCI)-Mayo Clinic summit webinars to be published. This webinar was hosted by the regular faculty, with Dr Anderson, a congenital heart disease expert from the Mayo Clinic, in attendance. He provides background on the lifetime management of repaired tetralogy of Fallot (rToF), followed by a clinical case (presented by Dr Engelbrecht) spanning 3 distinct phases in the patient's lifetime. The discussants are cardiology fellows from South African universities.

Objective: This manuscript, arising from the webinar series, summarises a multidisciplinary discussion on the lifelong management of rToF.

Design: Edited transcript of an expert webinar jointly hosted by SASCI and the Mayo Clinic faculty.

Case: The management of a female patient is discused in three stages at different age points of the patient's lifetime. The patient received a surgical rToF as a 1-year-old girl. She was then followed longitudinally into adulthood. At 19 years old, she was asymptomatic at presentation with severe pulmonary regurgitation (PR), progressive right ventricle (RV) remodelling, and borderline functional capacity. The discussion explored thresholds for intervention, imaging strategies where cardiac magnetic resonance imaging (CMR) access is limited, and surgical versus transcatheter valve replacement options. The patient ultimately underwent surgical pulmonary valve replacement (PVR) with a 27 mm bioprosthetic valve. At phase 3, she presented at the age of 29 with degeneration of her bioprosthetic valve. She was evaluated and received a transcatheter valve.

Key messages: (1) Understanding embryology and surgical history informs lifelong surveillance; (2) PR-driven RV remodelling is central to management; (3) objective imaging and functional markers (RV volumes, QRS, cardiopulmonary exercise test [CPET], arrhythmia burden) guide timing more reliably than symptoms alone; (4) procedural choice balances anatomy, concomitant lesions, device availability, and lifetime reintervention planning; and (5) lifelong exercise and structured follow-up are essential.

Online resource: Recorded SASCI fellows webinars (restricted to verified healthcare professionals) are available from: https://www.sasci.co.za/content/page/sasci-educational-videos1.

Keywords: tetralogy of Fallot, adult congenital heart disease, pulmonary regurgitation, right ventricle, pulmonary valve replacement, cardiology education, SASCI fellows webinar.

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INTRODUCTION

Moderators (Weich and Khan): Welcome, everyone. Tonight's topic is the lifetime management of adults with rToF. Adults now comprise most congenital heart disease survivors; most have had childhood repair and require lifelong surveillance. We'll focus on pathophysiology, what to monitor, imaging choices, indications, and timing for intervention – especially PVR – and how we decide between surgical and transcatheter options.

Why tetralogy of Fallot (ToF) looks the way it looks (Anderson)

Rather than memorising "ventricular septal defect (VSD), overriding aorta, pulmonary stenosis (PS), right ventricular hypertrophy", start with embryology: malalignment/rotation of the conotruncal septum shifts anteriorly, narrowing the right ventricular outflow tract (RVOT), creating a VSD and overriding

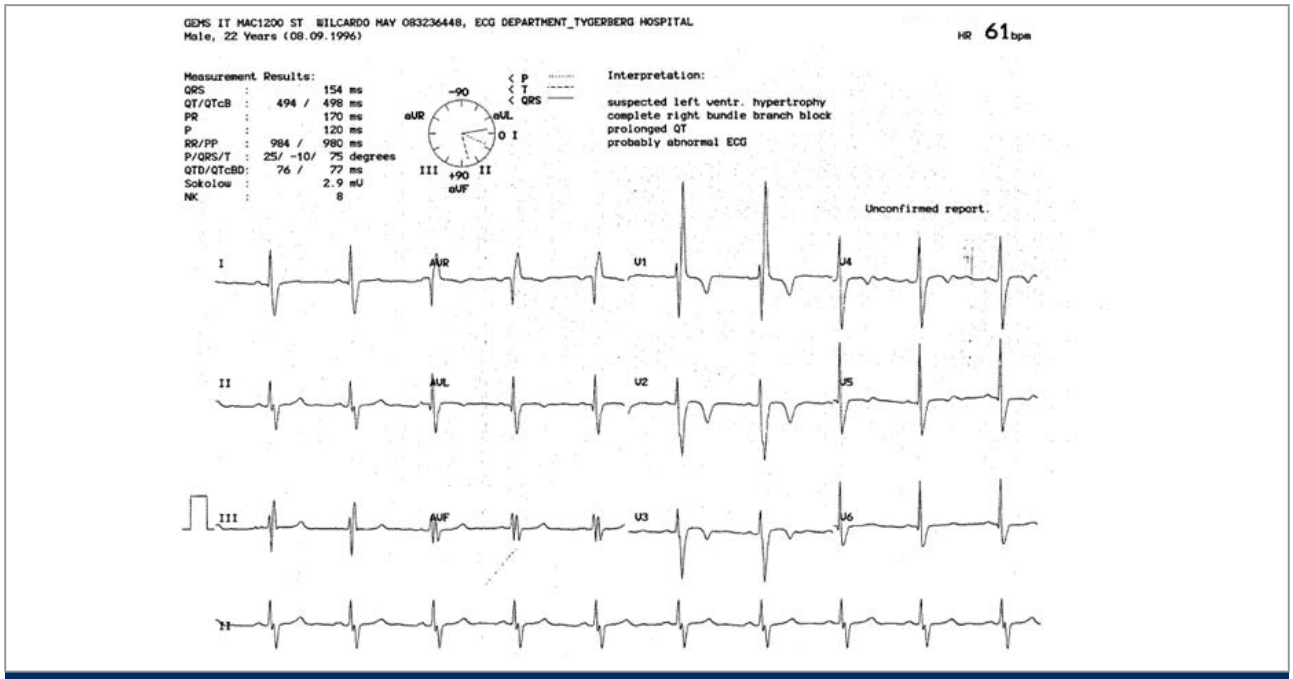

FIGURE 1: Electrocardiogram showing right bundle branch block and a QRS duration of 154 ms.

aorta. This spectrum explains variability – from classic ToF to ToF with pulmonary atresia – and why coronary variants crossing the RVOT matter surgically. Historically, ToF shaped paediatric cardiac surgery and modern transcatheter valves. The first transcatheter heart valve was deployed in the pulmonary position, and native/anastomotic RVOT anatomy still dictates strategy today.

Key pearl: Stenosis is poorly tolerated across the lifespan. Isolated PR is often well tolerated until RV remodelling accrues.

Residual lesions after childhood repair

- Common sequelae: Free PR after transannular patch, mixed PS/PR after valve-sparing repair, branch pulmonary artery (PA) issues at shunt sites, unexpected residual VSD, tricuspid regurgitation (TR) from RV dilation, left ventricular (LV) dysfunction from ventricular interaction, aortic root changes, and ventricular arrhythmias due to scar and dilation.
- RV remodelling matters: Progressive PR results in RV dilation leading to electromechanical dyssyncrony and LV dysfunction through ventritular interdependence. Our current practice focuses less on mortality (evidence limited) and more on how far remodelling can go, yet still reverse after PVR.
- Risk markers: LV systolic/diastolic dysfunction, non-sustained ventricular tachycardia (VT), QRS duration ≥ 180 ms, extensive RV scar, inducible VT.

CLINICAL CASE

Phase 1, age 1 year (Engelbrecht)

The patient was born with ToF and received a repair at Red Cross War Memorial Children's Hospital at the age of 1. She did very well after this and enjoyed a good quality of life throughout childhood.

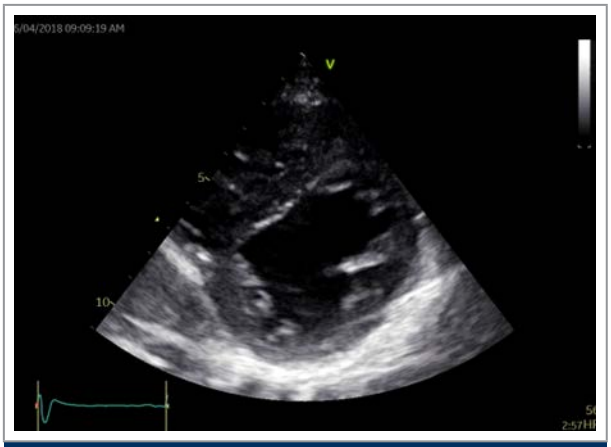

FIGURE 2: Echo showing D-shaping of the interventricular septum.

Phase 2, age 19 years

The patient presented for routine follow-up at her paediatrician. She continues to enjoy a good quality of life with no symptoms.

- Electrocardiogram: Sinus rhythm, right bundle branch block (RBBB), QRS 154 ms (Figure 1).
- Echo: Dilated proximal RVOT (45 mm), akinetic RVOT segment (likely secondary to a transannular/patch), paradoxical septal motion (Figure 2), severe PR with dense diastolic jet and brief PR pressure half-time (Figure 3),

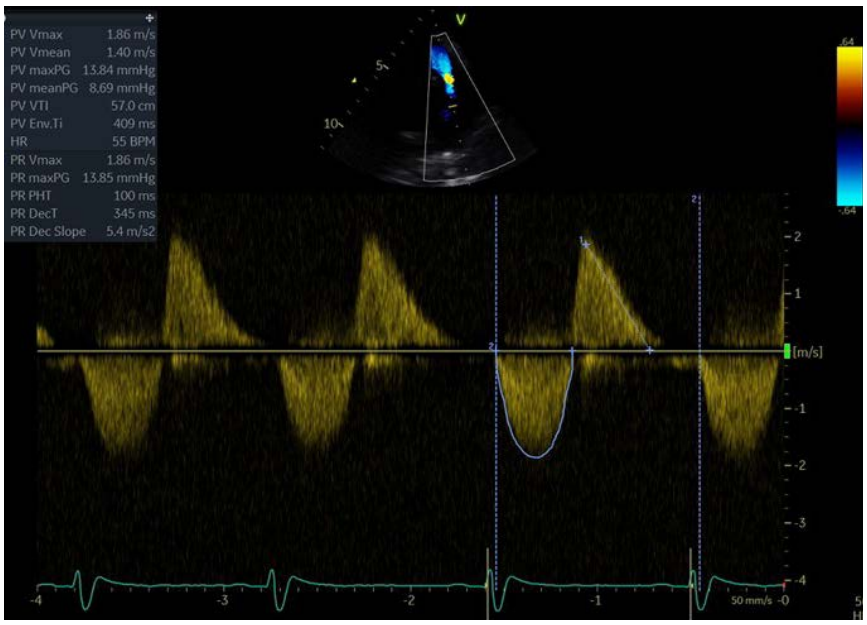

FIGURE 3: Continuous wave Doppler over the pulmonary valve showing a low systolic gradient over the valve (8 mmHg) but severe pulmonary regurgitation with a pressure half-time of 100 ms.

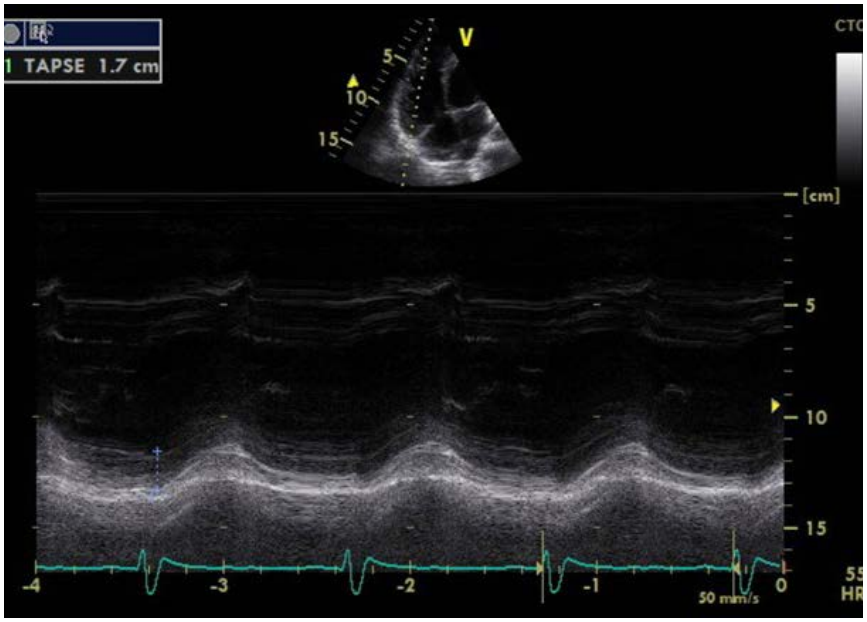

FIGURE 4: Echocardiography showing tricuspid annular plane systolic excursion (TAPSE) of 17 mm.

mildly reduced LV systolic function, tricuspid annular plane systolic excursion (TAPSE) of 17 mm, and S' 7 cm/s (suggesting mild RV systolic impairment in a volume-loaded RV) (Figures 4 and 5).

- CPET: VO_2 max 28 ml/kg/min (63% predicted), respiratory exchange ratio of 1.09 indicating adequate effort.
- CMR: Free PR, regurgitant fraction 58%, indexed RV end-diastolic volume (RVEDVi) 208 ml/m^2, indexed RV end-systolic volume (RVESVi) 97 ml/m^2, RV mildly to moderately impaired (right ventricular ejection fraction [RVEF] ≈ 40%), no branch PA stenosis. Apparent native annular remnant with post-stenotic main pulmonary artery (MPA) dilation – suggesting valve-sparing childhood repair.

Discussant 1: The central dilemma is an asymptomatic patient with severe PR and RV dilation/dysfunction. CPET shows reduced capacity, though not profoundly. Objective measures (volumes, function) guide us more than symptoms.

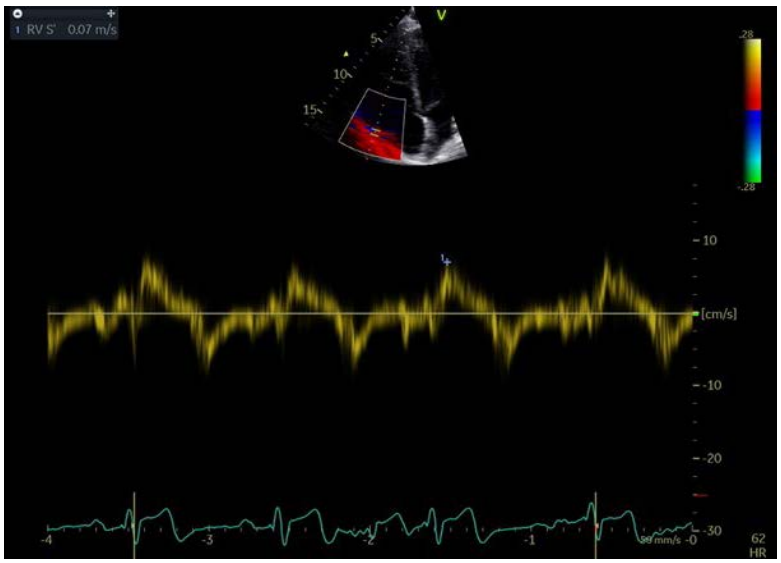

FIGURE 5: Echocardiography tissue Doppler showing a right ventricular S'- wave of 7 cm/s indicating right ventricle systolic dysfunction.

Discussant 2: CMR is the gold standard for RV volumes (notably the RVEDVi), but access is limited in many centres. Where CMR isn't feasible, serial echo (including RV:LV ratio on parasternal short-axis [PSAX]) and, if possible, computed tomography (CT) angiography with systolic/diastolic phases can help.

Weich (to panel): The QRS is 154 ms. Why mention it?

Anderson: It tracks with remodelling and arrhythmic risk. QRS ≥ 180 ms is a recognised risk threshold; watching the QRS trajectory helps frame timing even before symptoms declare themselves.

Echo pearls

On PSAX, RV is enlarged but not "2:1" versus LV. Septal flattening appears in diastole (volume load) rather than systole (pressure load). The Doppler profile shows early diastolic PR deceleration heading towards baseline – features compatible with significant PR. In volume-loaded physiology without restrictive RV features, the LV appears relatively preserved here.

Exercise and lifestyle

Aerobic conditioning improves outcomes across adult congenital heart disease (ACHD). We used to restrict exercise, but now encourage regular, structured exercise. Interpreting CPET needs context: 63% predicted in a sedentary person is not the same as 63% in a trained runner.

How do we decide on management at this stage?

Symptoms alone are unreliable in slow, insidious PR remodelling. Objective triggers discussed:

- Function: RV and/or LV systolic dysfunction (≥ mild-to-moderate) in ≥ moderate PR.
- Remodelling: RV enlargement (e.g. magnetic resonance imaging measured RVEDVi > 160 ml/m^2 and/or RV:LV end-diastolic volume ≈ 2:1).
- Haemodynamics: Significant RVOT obstruction (PS), elevated right ventricular systolic pressure (RVSP) (context-dependent) may indicate the need for surgery, although this was not a problem for the patient.
- Exercise capacity: Objective fall in exercise performance (e.g. CPET).
- Arrhythmias: non-sustained VT, inducible VT, or progressive QRS prolongation are potential indicators for intervention.

Choose surgery when concomitant work is needed (e.g. residual shunt, aortic work), complex arrhythmias requiring surgical ablation, small fixed surgical rings (e.g. small prosthetic heart valves) that will not accommodate dilation, high risk of coronary compression, or endocarditis. Choose a transcatheter for isolated RVOT disease, favourable anatomy, or high surgical risk – keeping in mind this is the first step in a lifetime sequence (valve-in-valve strategies, frame fracture, over-expansion planning).

Is there a place for medical therapy to prevent remodelling?

Evidence supports guideline-directed medical therapy for LV dysfunction in ACHD, but we lack convincing data that medical therapy alters RV volume-overload remodelling from PR. This may evolve with ongoing research.

What about pregnancy?

PR is generally tolerated in pregnancy; PS is more problematic. For a planned pregnancy with advanced remodelling, optimising physiology (e.g. timely PVR) may support maternal and foetal outcomes.

Case management

A multidisciplinary team weighed the substantial RVEDVi (208 ml/m^2), PR fraction 58%, QRS 154 ms, mildly reduced RVEF, and reduced CPET. Given the anatomy and local device availability, the team recommended a surgical bioprosthetic pulmonary valve replacement (27 mm Edwards PERIMOUNT [Edwards Lifesciences, Irvine, United States]). Post-operative echo showed a well-functioning valve with relief of PR.

Phase 3, age 29

At 29 years old, now a fitness instructor, the patient re-presented with reduced effort tolerance and near-syncope at high intensity.

- Echo: Mixed disease with severe PS (peak ~ 66 mmHg, mean ~ 37 mmHg) plus severe PR, markedly enlarged RV, and septal D-shaping. Invasive peak-to-peak RV-PA gradient 38 mmHg, RVSP ~ 65 mmHg.
- Doppler learning point (Discussant 1): PR backflow that equalises well before the next systole indicates severe regurgitation. In the presence of PS, derived RVSP largely reflects the stenotic gradient rather than true PA pressure.

Discussant 2 (strategy): Given prior surgeries and increasing risk with re-do surgery, transcatheter valve-in-valve is attractive when anatomy and frame size permits the use of locally available device options and when there is suitable expertise. In some settings, native anatomy can be tricky for transcatheter options, and device availability varies.

PRACTICAL TOOLBOX (FROM THE DISCUSSION)

Imaging when CMR is limited

- Serial transthoracic echo with RV:LV size ratio on PSAX, RVOT dimensions, PR Doppler contour, tricuspid regurgitation velocity context, and qualitative RV function.
- Consider CT angiography with systolic/diastolic reconstructions for volumetric estimates and RVOT geometry when CMR is unavailable. However, this will expose the patient to significant radiation.

Objective triggers used by the panel

- Worsening RV volumes (e.g. markedly elevated RVEDVi) or RV ≈ 2 × LV end-diastolic volume.
- Decline in RVEF and/or left ventricular ejection fraction, electromechanical dyssynchrony.
- CPET reduction versus expected training status.
- Arrhythmic markers: NSVT, progressive QRS prolongation, especially toward ≥ 180 ms.
- Development of PS (haemodynamically significant) or rising RV pressures.

Choosing surgical versus transcatheter PVR

- Surgery: Concomitant lesions (residual VSD, aorta), complex arrhythmias requiring surgical ablation, small nonfracturable ring, coronary compression risk, or active endocarditis (after acute phase).
- Transcatheter: Isolated RVOT disease, suitable landing zone (stented valve, conduit, or native RVOT, where dedicated systems are available), and a plan for lifetime valve-in-valve expansions.

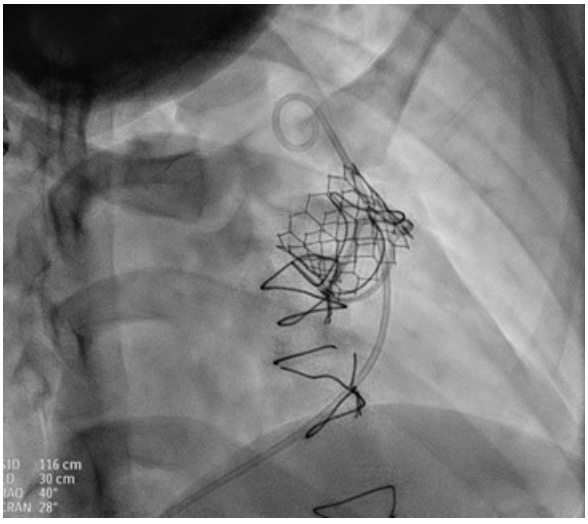

FIGURE 6: An Edwards SAPIEN 3 valve implanted within the degenerated bioprosthetic valve.

MANAGEMENT (Weich)

Because of the haemodynamic measurements and her symptoms, it was decided to perform a transcatheter pulmonary valve implant. Pre-operative CT scan excluded significant branch stenoses of her pulmonary arteries, and no hostile features at the landing zone. The Edwards PERIMOUNT valve has a true inner diameter of 25 mm, so we elected to implant a 29 mm Edwards SAPIEN 3 valve as recommended by the manufacturer. It is possible to perform a fracture of the bioprosthetic valve, but the very large fracture balloon required was unavailable. The SAPIEN 3 valve was implanted uneventfully under sedation, and she was discharged the next morning (Figure 6). At follow-up, she was asymptomatic and able to perform rigorous physical exercise.

Lifestyle and follow-up

- Encourage structured aerobic training across the lifespan.
- Regular electrocardiogram (QRS tracking), periodic CPET, and imaging keyed to prior findings.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

AUTHOR CONTRIBUTIONS

Case presentation and drafting: Engelbrecht. Conceptual content and editing: Anderson/Weich/Barsness/Holmes/Khan. All authors reviewed and approved the manuscript.