Fourth Decade After Repair of Tetralogy of Fallot
Taking Aim at Moving Targets

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This year marks the 60th year since the first successful intracardiac repair of tetralogy of Fallot (TOF). The repair, largely unchanged over decades, consists of ventricular septal defect closure and the relief of variable forms of right ventricular (RV) obstruction, usually with a transannular patch (TAP), and usually resulting in free pulmonary insufficiency (PI). Early in the experience with TOF repair, attention was on quantity of life—lifting the early dip in the survival curve. Palliative shunts were widely used to permit repair at a safer, older age. The dividends from a full relief of obstruction included excellent function for decades for patients formerly suffering from morbid or lethal disease. In this issue of Circulation, Cuypers and colleagues report findings of the third decennial follow-up of a cohort of early TOF repairs, the longest prospective study of this population to date. As such longitudinal evidence accrues, we are continually called on to reexamine both ends of the treatment timeline, although it is a wobbly yardstick that attempts to inform today’s best practices by measuring the sequelae of yesterday’s.

At the Front End

Early mortality after TOF repair is now very low, and the 10- to 20-year follow-up was encouraging. However, in the third postrepair decade, the Kaplan–Meier curve droops with progressive exercise intolerance, arrhythmia, right heart failure, and sudden death. Evidence points to ventricular scarring, chronic PI, RV hypertrophy, and declining left ventricular and RV performance as interrelated contributing factors. Some late risk may be modified at the front end, during infant repair. The progression of late sequelae also pushes the question of how to do a better job at the back end, with proactive strategies to preempt the onset of late morbidity.

At the Back End

Whereas the 40-year data presented today may questionably foresee the fate of today’s infants, they represent what is exactly relevant to today’s adults, who make up the drooping tail of the Kaplan–Meier curve and who account for a large proportion of adult congenital interventions today. Neither morbidity nor mortality plateaus in the fourth decade. Cuypers’ group shows a cumulative incidence of all events (death, cardiac reintervention, symptomatic arrhythmia, stroke, heart failure, endocarditis), continuing to worsen in the third decade, with event-free survival of only 25% at 40 years, consistent with other series.

Although PI itself is not an independent predictor of functional status, decompensated chronic volume load likely underlies the progression of ventricular dysfunction and associated late arrhythmia and mortality. In addition to fueling valve-sparing approaches at the front end, these findings underlie a controversial examination of timing for pulmonary valve replacement (PVR) in the adult with chronic PI. Does an asymptomatic patient with 25-year freedom from PVR count as a success or as a failure to intervene in time to forestall the decline that is already silently underway? In the absence of symptoms, heart failure, or arrhythmia, there is not a clear consensus on PVR indications. PVR is a treatment that can both cause and prevent a cardiac event. Is preventive maintenance a justified risk? Are there any patients who will never need PVR?

The criteria justifying PVR are reasonably moving in the direction of earlier intervention. A QRS duration of >180 milliseconds predicts ventricular tachycardia, and QRS duration...
stabilizes after PVR.13 Reasoned thresholds for PVR based on QRS prolongation have moved from 180 toward 140 milliseconds.12 Functional recovery of the RV after PVR is less likely if the preoperative RV end-diastolic volume is >170 mL/m², and that threshold is moving toward 150 mL/m².13 Exercise criteria, RV mass-to-volume ratio, and other metrics support lowering the threshold.13

The Rotterdam data report results of a conservative strategy, with PVR reserved for those with symptoms.14 The threshold for asymptomatic patients with PI has moved earlier in recent years, but long-term data on the effect of proactive intervention remain incomplete. Competing risks of a more aggressive approach to PVR include an uncertain benefit and the higher risk of reinterventions after earlier PVR. Overall, the mortality for PVR is low and getting lower, and durable improvement in symptoms and functional class after PVR has been shown, although a survival advantage has so far not.15,16 Further complicating the question are the unknowns about the long-term fate of catheter-based PVR. Clarity awaits long-term results of pulmonary valve replacement in repaired tetralogy of Fallot, who commonly disappear from surveillance presentation for treatment. This points to the importance of referral programs to identify earlier progression of silent disease.

The goal of health care is to optimize both quantity and quality of life for patients. Challenging evidence-based medicine is the fact that evidence commonly comes from groups, but medicine is applied to individuals. The challenge broadens when our evidence from yesterday’s groups is all we have to gauge what is best for today’s individual. As our experience with TOF repair enters a fifth decade, the steady progression of functional decline justifies earlier intervention at both ends of the treatment timeline, with encouraging early results that await the test of coming decades. Positive patient self-assessments in the face of declining physiological status should alert us to the need to further refine the threshold for PVR and to devise systematic surveillance systems to identify earlier progression of silent disease.

Disclosures

None.

References


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