low the transpulmonary gradient, in Fontan patients the primary risk factor for failure of the circulation is ventricular dysfunction.³

The Fontan conversion operation is successful in improving patient quality of life by restoring atrioventricular synchrony, which is so important to optimizing the engine and cardiac output.⁴ In nearly all of the patients requiring heart transplantation because of a failed Fontan circulation, the underlying etiology is pump, engine failure.⁵ This can be a combination of either systolic or diastolic dysfunction. Rychik notes, “Pulmonary vasoreactivity is likely just one of many determinants influencing pulmonary blood flow.”¹ The most important determinant is the engine and the cardiac output generated. In the current era where most single ventricle patients are

carefully staged to optimize pulmonary vascular resistance the importance of the engine remains paramount. Therapies to improve the quality of life in Fontan patients need to focus on optimizing the horsepower of the engine.

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Commentary: The Fontan: Propping up the push, the pull, the plumbing, and knowing when to fold

David P. Bichell, MD

Innovative plumbing configurations, refined over 5 decades, circumventing the “dispensable right ventricle,”¹ have delivered today’s patients with Fontan circulation a solid expectation for survival into adulthood.² Although surviving, even the “perfect Fontan” patient is long recognized to be plagued by late arrhythmia, chronic failure, gradually declining functional capacity, and premature mortality.³ Today, despite refinements, the Fontan works

COMPETING FACTORS AFFECTING FONTAN OUTCOME		
	SUCCESS PROMOTING FACTOR	MODIFIABLE CONTRAINDICATING FACTOR
PLUMBING	PASSIVE HYDRODYNAMICS	SUBOPTIMAL FONTAN ARCHITECTURE
		BRANCH PULMONARY STENOSIS
		SYSTEMIC VENOUS OBSTRUCTION
PUSH (PREPULMONARY)	LOW PULMONARY RESISTANCE	WASOREACTIVITY
		ACUTE PULMONARY COLLATERALS
		LYMPHATIC DYSFUNCTION
PULL (POSTPULMONARY)	EXTREMITY MUSCLE CONTRACTION	THROMBOEMBOLIC DISEASE
	NEGATIVE INTRATHORACIC PRESSURE	SEDENTARY LIFESTYLE
	DIASTOLIC HEALTH	ARTWAY DISEASE
RECOGNITION AND TREATMENT OF FAILURE	SYSTEMIC ATRIOVENTRICULAR VALVE DESCENT	DIASTOLIC DYSFUNCTION
	NORMAL SINUS RHYTHM	SALVAGE INEFFICIENCY
	ACHD SPECIALTY CLINIC FOLLOW UP (SURVEILLANCE ECHO, MRI, VOO)	SYSTEMIC DYSFUNCTION
	TIMELY MECHANICAL SUPPORT, TRANSPLANT	ARRHYTHMIA
		SOCIOECONOMIC DISTRESS/ACCESS TO CARE
		POOR TRANSITION TO ADULT CONSENTAL CARE
		SOCIOECONOMIC DISTRESS/ACCESS TO CARE
		EXTRACARDIAC ORGAN DECLINE

Deobstructing access to adult congenital care may contribute most to Fontan success.

CENTRAL MESSAGE

Despite Fontan optimization, failure ensues for many. Of modifiable factors, greatest impact may come from pre-emptive recognition of failure, a timely transition to specialized adult congenital care.

for some, not for all, and our predictive acumen remains limited. It is not clear who does or does not have a dispensable right ventricle.

Absent a subpulmonary ventricle, the competition for and against forward flow relies on the push (skeletal muscle pump), the pull, (respiratory muscle, ventricular diastolic health, systolic descent of the atrioventricular valve, rhythm

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COMPETING FACTORS AFFECTING FONTAN OUTCOME		
	SUCCESS-PROMOTING FACTOR	MODIFIABLE CONTRAVENING FACTOR
PLUMBING	PASSIVE HYDRODYNAMICS	SUBOPTIMAL FONTAN ARCHITECTURE
		BRANCH PULMONARY STENOSIS
		SYSTEMIC VENOUS OBSTRUCTION
	LOW PULMONARY RESISTANCE	VASOREACTIVITY
		AORTOPULMONARY COLLATERALS
		LYMPHATIC DERANGEMENT
		ENDOTHELIAL DYSFUNCTION
		THROMBOEMBOLIC DISEASE
PUSH (PREPULMONARY)	EXTREMITY MUSCLE CONTRACTION	SEDENTARY LIFESTYLE
PULL (POSTPULMONARY)	NEGATIVE INTRATHORACIC PRESSURE	SEDENTARY LIFESTYLE
		AIRWAY DISEASE
	DIASTOLIC HEALTH	DIASTOLIC DYSFUNCTION
	SYSTOLIC ATRIOVENTRICULAR VALVE DESCENT	VALVAR INSUFFICIENCY
	SYSTOLIC ATRIOVENTRICULAR VALVE DESCENT	SYSTOLIC DYSFUNCTION
RECOGNITION AND TREATMENT OF FAILURE	ACHD SPECIALTY CLINIC FOLLOW UP (SURVEILLANCE ECHO, MRI, VO ₂)	ARRHYTHMIA
		SOCIOECONOMIC DISTRESS/ACCESS TO CARE
	TIMELY MECHANICAL SUPPORT, TRANSPLANT	POOR TRANSITION TO ADULT CONGENITAL CARE
		SOCIOECONOMIC DISTRESS/ACCESS TO CARE
		EXTRACARDIAC ORGAN DECLINE

FIGURE 1. Deobstructing access to adult congenital care may contribute most to Fontan success. *ECHO*, Echocardiography; *MRI*, magnetic resonance imaging.

stability), and the preservation of kinetic energy through passive plumbing (Fontan geometry, modifiers of pulmonary resistance). Forces aligning to promote forward flow compete with those opposing it, only some of which are modifiable.

In this issue of the *Journal*, Rychik reviews results of the FUEL (Fontan Udenafil Exercise Longitudinal) trial, 400 adolescent and young adult Fontan patients randomized to receive or not a phosphodiesterase-5 inhibitor (udenafil), and frames the study's relevance to Fontan performance.⁴ The FUEL trial could not demonstrate a benefit in peak exercise capacity with vasodilator therapy. Rychik⁴ uses the FUEL trial as point of departure for an excellent discussion of elements, modifiable and not, that prop up forward flow in the Fontan circulation. Rychik suggests that pulmonary vasodilation may not have a major role in promoting Fontan flow, and that skeletal muscle contraction through submaximal activity such as walking may be a more important, underrecognized modifiable factor.

Although important to engage everything that could possibly help support Fontan circulation, the greatest impact on overall outcome may come from an earlier confession that even the optimized Fontan may not work,

earlier recognition of the signs of failure, earlier intervention, earlier mechanical assist and transplant. The Fontan destined for failure is commonly a young adult, in awkward or incomplete transition from pediatric care to adult follow-up, with shaky if any health care coverage, and multifactorial barriers to specialty adult congenital health care access. Too many Fontan failures are not recognized until arrhythmia, ascites, hepatic, or renal insufficiency advance risk or disqualify for transplant. Diagnostic advances are emerging to help see incipient failure before stepping over the cliff of compounding risk. Better recognition of when we are approaching the cliff (VO₂ [aerobic capacity] change, pulmonary vascular resistance change,⁵ strain pattern by serial magnetic resonance imaging,⁶ tissue Doppler imaging⁷) may prevent dangling from it by the fingertips (arrhythmia, cirrhosis, ascites). Despite new and encouraging ways to detect early decline, failed access to specialty surveillance is a blindfold on approach to the risk cliff and a missed opportunity. The next high impact refinements to improve Fontan outcome may rest in the challenging area of socioeconomic change and the restructuring of transitional health care.

Submaximal activity no doubt improves Fontan circulation and should be promoted. Systemic changes in access

to and the provision of specialized adult congenital health-care may be a bigger modifiable factor promoting longevity and functional outcome (Figure 1).

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