further research.^{5,6} Although the wisdom of Diogenes *Solvitur ambulando*, or the problem is solved by practical experimentation, is relevant today, perhaps equally important is to apply “scientific methodology” first introduced by Aristotle, another ancient Greek philosopher (384–322 BC). Well-designed, multicenter, randomized, double-blind, and controlled studies examining the determinants of pulmonary blood flow and their effect in optimizing Fontan circulation will afford the medical community the benefit of pushing the frontiers in this field with great benefits for these patients.

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Check for updates

Commentary: It's the engine, not the fuel

Carl L. Backer, MD

Rychik¹ has provided an insightful analysis of the somewhat disappointing results from the very well-conducted Fontan Udenafil Exercise Longitudinal (FUEL) study. He acknowledges this study to be a true tour de force, with hundreds of patients enrolled across multiple institutions; the largest clinical trial of its kind ever undertaken in congenital heart care. Part of the disappointment of this trial is that health care providers managing the care of Fontan patients are clearly hungry to discover and employ new, effective strategies to improve the quality of life of these patients. Rychik¹ notes that although treatment with udenafil was not associated with an improvement in oxygen consumption at peak exercise, there was some improvement in exercise performance at submaximal activities like walking. This activity may actually be more important than the findings at peak exercise and probably merits further investigation.

Carl L. Backer, MD

CENTRAL MESSAGE

Decreasing pulmonary vascular resistance may provide marginal improvement for Fontan patients, but it is the strength of the engine in the circulation that is the most important factor for determining pulmonary blood flow.

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In addition to pulmonary vasoreactivity, I believe we actually do understand the relative magnitude of factors contributing to pulmonary blood flow in the Fontan circulation. The primary driver is the engine—the cardiac output generated by the single ventricle, which will not be affected by FUEL. My experience with patients requiring surgical therapy for failing Fontans has been that in nearly all circumstances it is the engine that is the source of Fontan failure.² Medical manipulation of the pulmonary vascular resistance as demonstrated by the FUEL trial had a minimal effect. It is the pump, the single ventricle, the engine that is the primary driver of pulmonary blood flow. No matter how

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
LOW PULMONARY RESISTANCE		VASOREACTIVITY
		ACUTE PULMONARY COLLATERALS
		LYMPHATIC DYSFUNCTION
PUSH (PREPULMONARY)	EXTREMITY MUSCLE CONTRACTION	SEDENTARY LIFESTYLE
PULL (POSTPULMONARY)	NEGATIVE INTRATHORACIC PRESSURE	SEDENTARY LIFESTYLE
		ARTWAY DISEASE
RECOGNITION AND TREATMENT OF FAILURE	DIASTOLIC HEALTH	DIASTOLIC DYSFUNCTION
		SALIVARY INSUFFICIENCY
TIMELY MECHANICAL SUPPORT, TRANSPLANT	SYSTOLIC ATRIOVENTRICULAR VALVE DESCENT	SYSTOLIC DYSFUNCTION
ACHD SPECIALTY CLINIC FOLLOW UP (SURVEILLANCE ECHO, MRI, VOO)	NORMAL SINUS RHYTHM	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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