further research. 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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**Commentary: It’s the engine, not the fuel**

Carl L. Backer, MD

Rychik 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 hun-
dreds 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.
Rychik 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 ac-
tivity may actually be more important than the findings at
peak exercise and probably merits further investigation.

In addition to pulmonary vasoreactivity, I believe we
actually do understand the relative magnitude of factors
contributing to pulmonary blood flow in the Fontan circula-
tion. 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 cir-
cumstances it is the engine that is the source of Fontan fail-
ure. 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.2

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.2 In nearly all of the patients requiring heart transplantation because of a failed Fontan circulation, the underlying etiology is pump, engine failure.2 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 center stage to optimize the horsepower of the engine.

References

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,” 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 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.

CENTRAL MESSAGE

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

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