

Development of a Fontan Survivorship and Advanced Therapy Program: A Journey from Quality Improvement to Action

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Background: Survival after the Fontan operation for patients with functional single ventricle congenital heart disease has improved in the last three decades. However, survivors are at risk for a wide variety of cardiac and non-cardiac complications that can impact late morbidity, mortality, and quality of life. These complications include ventricular dysfunction, exercise intolerance, protein losing enteropathy, plastic bronchitis, liver disease, impaired somatic growth, endocrine dysregulation, and neurocognitive impairment among others. Currently there are no consensus statements or guidelines on optimal surveillance, risk stratification, and management for this steadily growing population. An evolving strategy is the development of a specialized multidisciplinary program led by providers dedicated to the care of patients with Fontan physiology¹; however, the outcomes of this strategy in improving quality of life and morbidity in this population are unknown. The aim of this study is to review the preliminary outcomes of our novel multi-disciplinary Fontan Survivorship & Advanced Therapy Program as a strategy to manage patients with high-risk or failing Fontan physiology.

Methods: In 2014, we developed an evidence-based quality improvement protocol to standardize the assessment and management of Fontan patients.^{2,3} The goal of the protocol was to improve identification of Fontan-related complications, as well as asymptomatic Fontan patients who are high-risk for morbidities or Fontan failure. This protocol heightened awareness of risk factors impacting morbidity, need for advanced management strategies, and collaboration with non-cardiac specialists. To address these needs we created the Fontan Survivorship & Advanced Therapy Program. This novel, multidisciplinary team includes cardiology physicians and advanced practice nurses, as well as subspecialists from gastroenterology, hepatology, endocrinology, pulmonology, nutrition, psychology, and palliative care. The team meets monthly providing both inpatient and outpatient consultation for Fontan patients referred for symptoms or abnormal testing results. We reviewed the first eight months of clinical activity including our inpatient and outpatient assessments from 1/1/16 to 9/1/16. Patient demographic, diagnostic, laboratory, imaging, and hemodynamic data were collected and reviewed. Standard descriptive statistics were utilized to characterize clinical data and to depict the initial outcomes of the clinic.

Results: Since inception of our Fontan team in January of 2016, 13 patients have been referred for assessment including 11 outpatient and 2 inpatient consultations. Indications for referral to the Fontan Survivorship & Advanced Therapy Program have included protein-losing enteropathy (6), ventricular dysfunction (4), plastic bronchitis (1), hepatic nodule (1), and pre-operative cardiac clearance for high-risk non-cardiac surgery (1). NYHA classification at time of initial FACT clinic evaluation has been NYHA I in 0, II in 11, III in 2 and IV in 0 and ACC/AHA Heart Failure Stage has been A in 0, B in 4, C in 9 and D in 0. Standardized assessment including lab, imaging, nutrition and psychology was completed on all 13 patients and revealed average O₂ saturation of 89% (81-96%; Z=5.4), NT-pro-BNP 484 (41-1030; Z=328), and albumin of 4 (2.4-5.1 Z=0.8). One patient is actively under assessment of a concerning hepatic nodule and 6 others are seen concurrently by pulmonology and 6 by gastroenterology and 6 by psychology.

Conclusion: A programmatic quality assessment of Fontan management within our heart center demonstrated the need for a specialized multidisciplinary referral clinic for Fontan patients with complications and/or high-risk features. Our initiation of this clinic has identified 13 patients with significant morbidities that required additional medical interventions, advanced imaging, or continued care by dedicated non-cardiac subspecialists. This monitoring program is a first step in identifying patients in need of a higher level of care, advanced interventions and hopefully to allow timely interventions for Fontan morbidities.

Refs:

1. New Concepts: development of a survivorship programme for patients with a functionally univentricular heart. Goldberg D et al. *Cardiology in the Young* 2011, 21(2) 77-79
2. Quality Indicators for Adults with Congenital Heart Disease: A Single Program Assessment of Current Practice, Nancy Rudd, APNP, et al; *MWPCS Annual Meeting 2014*
3. Ginde S, Rudd N, Katzmark L, Sowinski J, Bonnell E, Earing M, Quality Indicators for Adults with Congenital Heart Disease: A Single Program Assessment of Current Practice, 24th Annual International Symposium on Congenital Heart Disease in the Adult, Cincinnati, OH, 06/2014