

# Fontan Executive Summary

Physician Owners: Dr. Vaikom House & Dr. Cramer

## PRIMARY OBJECTIVE

Develop a pathway for managing patients post-operatively after elective Fontan procedure in the PICU and inpatient floors and effectively transition to an outpatient setting

## CLINICAL CARE GUIDELINES

### Intended for patients:

- All patients referred for an elective Fontan procedure

### Not intended for patients with:

- Fontan revision
- Patients who remain intubated on transfer from OR to ICU following Fontan procedure

## CLINICAL MANAGEMENT

- All elective Fontans, extubated in the OR, stable off pressors > 4 hours at 0700 AM rounds on post-operative day (POD) 1 will be included
- Patients will be initiated on the following regimen on POD 1
  - F** – IV FUROSEMIDE 1mg/kg every 8h on POD 1. Aldactazide 1mg/kg PO every 12 hours to be initiated when taking enteral liquids. Can transition to PO tid furosemide after chest tubes are removed and will be discharged on po tid Lasix.
  - O** – Do not wean OXYGEN below 0.5 L/min via nasal cannula until chest tubes are removed. PT/OT should be consulted to assist with ambulation, okay for patient to be off oxygen while ambulating
  - N** – Initiate ENALAPRIL 0.05mg/kg/dose every 12h when taking PO fluids. Dose to be titrated up as needed for hypertension
  - T** – CHEST TUBES will be removed when drainage is < 2ml/kg/day per side or < 7 ml/kg/day for a single chest tube
  - A** – ANTICOAGULATION Patients will be placed on a heparin drip POD 0 (when 4 hours post op and without excessive bleeding), maintain therapeutic Anti-Xa until they are freely ambulating. Start aspirin on POD 1 when taking PO.
  - N** – NUTRITION and fluids. On POD 1-3, when PO is initiated, restrict total fluids to 80% of maintenance. Then increase to maintenance fluids until chest tube is removed. Maintain on a fat free diet (< 3 gms of fat) for 3 days, then increase to < 30% of total calories from fat until 4 weeks post op.

## RATIONALE

- **Safety:** Will be maintained by close communication between ICU physician, ICU cardiologist, CT surgeon, CT surgery advanced practice providers, inpatient Cardiologist and inpatient Cardiology nurse
- **Quality:** Will be improved by reducing practice variation
- **Cost:** Will be reduced by reducing practice variation in treatment which can lead to potential increased LOS, morbidity and readmissions
- **Engagement:** Is created and supported by involvement of providers across the continuum of care that evaluate and treat cardiology patients
- **Patient/Family Satisfaction:** Shall be improved by providing the highest quality care based on established guidelines and the latest evidence available in the literature

## IMPLEMENTATION ITEMS/ SUPPORTING DOCUMENTS

- CARD Fontan Post-op Day 1 Order Set

## METRICS PLAN

- Post-op Fontan pathway order set utilization rate
- ICU LOS
- Monitor chest tube days and reinsertion rate of chest tube
- Hospital LOS
- Readmissions within 30 days

## TEAM MEMBERS

Dr. James Hammel (Cardiothoracic Surgeon)  
 Dr. Vaikom House & Dr. Jonathan Cramer (Cardiology physicians)  
 Amy Kenkel, Valoree Stuhr, Holly Baynes, Kristy Cook (Cardiothoracic surgery Nurse Practitioners)  
 Venus Anderson RN (Cardiology service nurse)  
 Hannah Brewer RN (PICU)  
 Cynthia Wilson (Dietician)  
 Josh Werner (Dietician)  
 Janet Vetter (IT)  
 Melisa Paradis RN (Performance Improvement)

## EVIDENCE

1. A Multicenter, Randomized Trial Comparing Heparin/Warfarin and Acetylsalicylic Acid as Primary Thromboprophylaxis for 2 Years After the Fontan Procedure in Children  
 Paul Monagle, MD, MSC, MBBS,\* Andrew Cochrane, MD,\* Robin Roberts, MSC, Cedric Manlhiot, BSC,‡ Robert Weintraub, MBBS,\* Barbara Szechtman, BA, Marina Hughes, DPHIL,§ Maureen Andrew, MD,‡ Brian W. McCrindle, MD, MPH, for the Fontan Anticoagulation Study Group Melbourne, Victoria, Australia; and Toronto and Hamilton, Ontario, Canada
2. A Medical Strategy to Reduce Persistent Chest Tube Drainage After the Fontan Operation Joseph R. Cava, MD, PhD,a\*, Sarah M. Bevandic, RNa, Michelle M. Steltzer, CPNPa, and James S. Tweddell, MDb
3. Endothelial Function Following the Fontan Operation William T. Mahle, MD, Kathy Todd, RN, and Derek A. Fyfe, MD, PhD
4. Factors Associated With Thrombotic Complications After the Fontan Procedure A Secondary Analysis of a Multicenter, Randomized Trial of Primary Thromboprophylaxis for 2 Years After the Fontan Procedure Brian W. McCrindle, MD, MPH,\* Cedric Manlhiot, BSC,\* Andrew Cochrane, MD,† Robin Roberts, MSC,‡ Marina Hughes, DPHIL,† Barbara Szechtman, BA,‡ Robert Weintraub, MBBS,† Maureen Andrew, MD,\* Paul Monagle, MD, MSC, MBBS,† for the Fontan Anticoagulation Study Group Toronto and Hamilton, Ontario, Canada; and Melbourne, Australia
5. Factors Associated With Prolonged Recovery After the Fontan Operation Joshua W. Salvin, MD, MPH; Mark A. Scheurer, MD; Peter C. Laussen, MBBS; John E. Mayer, Jr, MD; Pedro J. del Nido, MD; Frank A. Pigula, MD; Emile A. Bacha, MD; Ravi R. Thiagarajan, MBBS, MPH
6. Factors Related to Pleural Effusions After Fontan Procedure in the Era of Fenestration Raymond T. Fedderly, MD; Beth N. Whitstone, RN; Stephanie J. Frisbee, MSc; James S. Tweddell, MD; S. Bert Litwin, MD
7. Fenestration Improves Clinical Outcome of the Fontan Procedure. A Prospective, Randomized Study Fontan Palliation in the Modern Era: Factors Impacting Mortality and Morbidity James S. Tweddell, MD, Matthew Nersesian, BS, Kathleen A. Mussatto, PhD, RN, Melodee Nugent, MA, Pippa Simpson, PhD, Michael E. Mitchell, MD, Nancy S. Ghanayem, MD, Andrew N. Pelech, MD, Rammohan Marla, MD, and George M. Hoffman, MD Division of Cardiothoracic Surgery, Department of Surgery, Sections of Pediatric Cardiology and Critical Care, Departments of Pediatrics and Quantitative Health Sciences, Medical College of Wisconsin and Herma Heart Center, Children's Hospital of Wisconsin, Milwaukee, Wisconsin Matthew S. Lemler, MD; William A. Scott, MD; Steven R. Leonard, MD; Daniel Stromberg, MD; Claudio Ramaciotti, MD
8. Fontan Physiology Revisited Matthew Jolley, MD,\*† Steven D. Colan, MD,† Jonathan Rhodes, MD,† and James DiNardo, MD\*
9. Management of the Failing Fontan Fabrizio DeRita, a David Crossland, b Massimo Griselli, a and Asif Hasana

10. The Fontan Patient: Inconsistencies in Medication Therapy Across Seven Pediatric Heart Network Centers Page A. W. Anderson • Roger E. Breitbart • Brian W. McCrindle • Lynn A. Sleeper • Andrew M. Atz • Daphne T. Hsu • Minmin Lu • Renee Margossian • Richard V. Williams
11. Reduced pleural drainage, length of stay, and readmissions using a modified Fontan management protocol Nancy A. Pike, PhD,a,b Carol A. Okuhara, MN,a Joy Toyama, MS,b Barbara P. Gross, MSN,a Winfield J. Wells, MD,a,c and Vaughn A. Starnes, MDa,c Increased platelet reactivity and significant changes in coagulation markers after cavopulmonary connection H B Ravn, V E Hjortdal, E V Stenbog, K Emmertsen, O Kromann, J Pedersen, K E Sorensen
12. A Defined Management Strategy Improves Early Outcomes After the Fontan Procedure: The Portland Protocol Rachel E. Sunstrom, PA-C, Ashok Muralidaran, MD, Rabin Gerrah, MD, Richard D. Reed, PA-C, Milon K. Good, MPH, Laurie R. Armsby, MD, Andrew J. Rekito, MS, M. Mujeeb Zubair, MD, and Stephen M. Langley, MD, FRCS (CTh) Divisions of Pediatric and Congenital Cardiac Surgery, Pediatric Cardiology, and Neurologic Surgery, Doernbecher Children's Hospital, Oregon Health and Science University, Portland, Oregon
13. Management of the postoperative Fontan patient Deborah R. Gersony\*, Welton M. Gersony
14. Quality-Cost Relationship in Congenital Heart Surgery Sara K. Pasquali, MD, MHS, Jeffrey P. Jacobs, MD, Edward L. Bove, MD, J. William Gaynor, MD, Xia He, MS, Michael G. Gaies, MD, MPH, Jennifer C. Hirsch-Romano, MD, MS, John E. Mayer, MD, Eric D. Peterson, MD, MPH, Nelangi M. Pinto, MD, MS, Samir S. Shah, MD, MSCE, Matt Hall, PhD, and Marshall L. Jacobs, MD Department of Pediatrics and Communicable Diseases, C.S. Mott Children's Hospital, Ann Arbor, Michigan; Department of Surgery, Johns Hopkins University School of Medicine, Baltimore, Maryland; Department of Cardiac Surgery, University of Michigan Medical School, Ann Arbor, Michigan; Department of Surgery, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania; Duke Clinical Research Institute, Duke University School of Medicine, Durham, North Carolina; Department of Cardiovascular Surgery, Boston Children's Hospital, Boston, Massachusetts; Department of Pediatrics, Primary Children's Hospital, Salt Lake City, Utah; Department of Pediatrics, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio; Children's Hospital Association, Overland Park, Kansas
15. Thromboembolism and the Role of Anticoagulation in the Fontan Patient M. L. Jacobs Æ K. K. Pourmoghadam
16. Strategies for thromboprophylaxis in Fontan circulation: a meta-analysis Tarek Alsaied,<sup>1</sup> Said Alsidawi,<sup>2</sup> Catherine C Allen,<sup>1</sup> Jenna Faircloth,<sup>3</sup> Joseph S Palumbo,<sup>4</sup> Gruschen R Veldtman
17. Thrombotic Complications and Thromboprophylaxis Across All Three Stages of Single Ventricle Heart Palliation Cedric Manlhiot, BSc<sup>1</sup>, Leonardo R. Brand~ao, MD<sup>2</sup>, Judith Kwok, BSc<sup>1</sup>, Stefan Kegel, MD<sup>1</sup>, Ines B. Menjak, MD<sup>1</sup>, Caitlin L. Carew, MSc<sup>1</sup>, Anthony K. Chan, MD<sup>3</sup>, Steven M. Schwartz, MD<sup>1</sup>, V. Ben Sivarajan, MD<sup>1</sup>, Christopher A. Caldarone, MD<sup>1</sup>, Glen S. Van Arsdell, MD<sup>1</sup>, and Brian W. McCrindle, MD, MPH
18. Variation in Outcomes for Benchmark Operations: An Analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database Jeffrey Phillip Jacobs, MD, Sean M. O'Brien, PhD, Sara K. Pasquali, MD, Marshall Lewis Jacobs, MD, Francois G. Lacour-Gayet, MD, Christo I. Tchervenkov, MD, Erle H. Austin III, MD, Christian Pizarro, MD, Kamal K. Pourmoghadam, MD, Frank G. Scholl, MD, Karl F. Welke, MD, and Constantine Mavroudis, MD The Congenital Heart Institute of Florida (CHIF), All Children's Hospital, University of South Florida College of Medicine, Cardiac Surgical Associates of Florida (CSAoF), Saint Petersburg and Tampa, Florida; Duke University School of Medicine and Duke Clinical Research Institute, Duke University Medical Center, Durham, North Carolina; Cleveland Clinic Lerner School of Medicine, Cleveland, Ohio; Children's Hospital at Montefiore, New York, New York; Montreal Children's Hospital, Montreal, Quebec, Canada; Kosair Children's Hospital, University of Louisville, Louisville, Kentucky; Alfred I. duPont Hospital for Children, Wilmington, Delaware; University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma; Joe DiMaggio Children's Hospital, Hollywood, Florida; and Seattle Children's Hospital, Seattle, Washington