Diagnostic Cardiac Caths - Pre-Glenn and Pre-Fontan: What are we looking for?

Matthew J. Gillespie MD, FSCAI
The Children’s Hospital of Philadelphia
SCAI Fall Fellows Course
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Presenter Disclosure Information

Matthew J. Gillespie, MD
No Disclosures
Goals of Pre-Glenn Cath?

Set the stage for “smooth sailing” intraoperatively and postoperatively
1. Hemodynamic assessment
   • Cardiac output, Qp:Qs
   • PA pressure / PVR
   • Ventricular End Diastolic Pressure
   • Systolic pressure gradients

2. Angiography
   • PA anatomy
   • Pulmonary venous anatomy
   • Aortic arch reconstruction
Stage 2 (Glenn or HemiFontan) is generally considered a "volume unloading" procedure resulting in completely passive flow through the lungs.
Not all “Stage 1’s” are equal..

• Approach to Pre-Glenn catheterization varies on type of stage 1

• ANATOMY DICTATES approach to access and angiography
  1. Norwood + BTS
  2. Norwood + RV-PA conduit (Sano modification)
  3. Hybrid approach (PA bands, PDA stent, ASD creation)
Assumed VO2 = 150 ml/min/m2
Hb = 15 g/dL
Stage 1 RMBTS

Assumed VO2 = 150 ml/min/m2
Hb = 15 g/dL

Qp = 3.1 L/min/m2
Qs = 1.8 L/min/m2

PVR = 2.9
SVR = 30

Qp:Qs = 1.7
Pre-Glenn Angiography: Stage 1 RMBTS

• What are we looking for?
  – Pulmonary venous anatomy
  – Pulmonary arterial distortion requiring “fixing” in the OR
  – Aortic arch obstruction
  – Native-to-neo aortic anastomosis
    • Coronaries? (sometimes)
  – AV valve regurgitation
  – Aortic valve regurgitation
  – LSVC-to-coronary sinus (or other potential source for systemic venous decompression to pulmonary veins)
Pre-Glenn Angiography: pulmonary veins
Pre-Glenn Angiography: RMBTS

“good one”
Pre-Glenn Angiography: RMBTS

“not so good one”
Pre-Glenn Angiography: RMBTS

“not so good one” post stent placement
Pre-Glenn Angiography: Innominate vein

Rule out LSVC
Pre-Glenn Angiography: Innominate vein

Rule out LSVC...
Pre-Glenn Angiography: Aortagram

Rule out / treat coarctation
Pre-Glenn Angiography: Aortagram

Post Balloon angioplasty
Hemodynamics & Pre-Glenn Cath: Norwood + RV-PA conduit

Assumed VO2 = 150 ml/min/m2
Hgb = 15g/dL
Hemodynamics & Pre-Glenn Cath: Norwood + RV-PA conduit

Assumed VO2 = 150 ml/min/m2
Hgb = 15g/dL

Qp = 2.5 L/min/m2
Qs = 4.1 L/min/m2

Qp:Qs = 0.6

PVR = 2.4
SVR = 11.2
Pre-Glenn Angiography:
Stage 1 RV-PA conduit

• What are we looking for?
  – Pulmonary venous anatomy
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  – AV valve regurgitation
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  – LSVC-to-coronary sinus (or other potential source for systemic venous decompression from above)
Angiography Pre-Glenn: Norwood + RV-PA conduit

RV gram
Angiography Pre-Glenn: Norwood + RV-PA conduit

RV-PA conduit-o-gram
Hemodynamics & Pre Comprehensive stage 2: Hybrid Stage 1

Typical CHOP hybrid patient

5 month F with HLHS, scimitar syndrome s/p Hybrid; hemodynamic cath to evaluate transplant candidacy

GETA, 40% FiO2 (MRI performed under same conditions)
7.47/4.4/28/31/+6.4, hgb 12.3
aVO2 170 mL/min/m2

By MRI:
- RPA = 1.4 L/min/m2
- LPA = 2.2 L/min/m2
- PVRI(R) = 22 IVU
- PVRI(L) = 5 IVU
- PVRI(T) = 4.1 IVU

No interventions
Complications: transient catheter-induced heart block with hypotension requiring brief resuscitation
Angiography Pre Comp Stg 2: Hybrid Stage 1

Ductal stent and PA bands
Pre-Glenn ("stage 2") Catheterization Summary

• “Set the table” for operative plan and postoperative management
  – Identify “suitability” for passive blood flow into lungs
  – Identify and treat anatomic issues
    • Coarctation of the aorta
    • LSVC-to-CS or other decompressing veins
    • Etc.
Pre-Fontan diagnostic catheterization
Goals of Pre-Fontan Cath?

1. Hemodynamic assessment
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   • PA pressure / PVR
   • Ventricular End Diastolic Pressure
   • Systolic pressure gradients

2. Angiography
   • PA anatomy
   • Pulmonary venous anatomy
   • Aortic arch reconstruction
Indications for Pre-Fontan Cath?

Diagnostic Assessment Before Fontan Bidire

Are Not

Pamela S.
William T
Philadelphia

What is Before a

Puja Banka ·
John E. Mayo
Tal Geva · D.

Is Routine Cardiac Catheterization Necessary in the Management of Patients with Single Ventricles Across Staged Fontan Reconstruction? No!

M.A. Fogel1,2

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Goals of Pre-Fontan Cath?

1. Hemodynamic assessment
   - Cardiac output, Qp:Qs
   - PA pressure / PVR
   - End Diastolic Pressure
   - Systolic pressure gradients

2. Angiography
   - PA anatomy
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Pre-Fontan catheterization: Hemodynamics

Assumed VO2 = 150 ml/min/m2
Hgb = 15 g/dl
Pre-Fontan catheterization: Hemodynamics

Assumed VO2 = 150 ml/min/m2
Hgb = 15g/dl

Qp:Qs = 0.5
Qp = 2.3 L/min/m2
Qs = 4.8 L/min/m2
PVR = ??

Pre-Fontan catheterization: Angiography

- SVC angiogram
- PA anatomy
- Transit time (AVMs)
- Competitive flow?
Pre-Fontan catheterization: Angiography

- Aortagram
  - AI?
  - Arch Obstruction?
  - Collateral flow??
Pre-Fontan catheterization: Angiography

Coronary picture
(native ascending aorta picture)
Systemic-to-pulmonary artery collaterals: Friend or foe?

Figure 3. Vascular sites of origin of all aortopulmonary collateral vessels identified in this study. Int. = internal.
What are SPCs?

• Connections between systemic arteries and pulmonary circulation

• Typically arise from DAO, subclavian, bronchials, and intercostals

• Unlike typical bronchials, they connect to the terminal respiratory unit (distal segments pulmonary vascular tree)
What causes SPC to form?

• Etiology in single ventricle patients?
  – Hypoxia ✔
  – Decreased pulmonary blood flow ✔
  – non-pulsatile pulmonary blood flow ✔
  – Postoperative inflammation (post thoracotomy) ✔
  – Absence of Hepatic factor ✔
Are there “positive” effects of SPCs on Single Ventricle Patients?

• Augment Qp to under perfused lung segments and therefore increase systemic arterial $O_2$ saturation

• Inhibit pulmonary AVMs by providing lungs with “hepatic factor”
“Negative” effects of SPCs on Single Ventricle Patients

- Reduces “effective” pulmonary blood flow
- Increase PA pressure and therefore systemic venous pressure, decrease in circulatory efficiency
  - Low cardiac output, effusions, hepatic congestion, peripheral edema, hemoptysis
- Volume load single ventricle
  - AVVR and global dysfunction over time
Do SPCs adversely effect Single Ventricle Outcomes?

**YES**

**NO**
### Previous Studies: Summary

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Where does this leave us?

Editorial

The Argument for Aggressive Coiling of Aortopulmonary Collaterals in Single Ventricle Patients

Herbert J. Stern, MD, FACC, FSCAI

Aortopulmonary collaterals are present in up to 80% of single ventricle anatomy patients undergoing pre-Fontan catheterization. There is generally no consensus as to their impact in the mid-long term course of these patients. We present arguments that aggressive coiling of these vessels is warranted to prevent deterioration of ventricular function long-term. Non-invasive modalities for measuring APC are needed. Multi-institutional, prospective, randomized, double blind studies are also needed to confirm our hypotheses. © 2009 Wiley-Liss, Inc.

Key words: coil occlusion; aortopulmonary collaterals; single ventricle
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Finally an answer……? 

• Maybe if we increase the “N” and look retrospectively across centers we’ll find an answer?

• Use the PHN Fontan Cross-Sectional data to look for outcome differences

• Methods
  – Impressive unprecedented, multicenter look into Fontan outcomes
  – 539 patients
    • 80 (15%) had “coils” = coil group
    • 459 (85%) = “no coils”
    • Hospital LOS = primary outcome
Frequency of coil embolization of APC vessels among centers. Center-specific proportions of subjects with APC coils ($P < .001$). Each bar represents 1 of the 7 centers in decreasing order of center sample size, with the number of subjects enrolled from the center listed atop the bar.
Figure 2

No difference in LOS

Post-Fontan Hospital LOS after adjustment for fenestration and year of Fontan. Proportion of subjects in the hospital versus days after Fontan for each group after adjusting for surgical fenestration and year of Fontan. Hazard ratio for remaining in the hospital for subjects in the coil versus no-coil groups is 0.91, 95% CI 0.70 to 1.18, P = .48.
Valvular and Congenital Heart Disease

Practice variability and outcomes of coil embolization of aortopulmonary collaterals before Fontan completion: A report from the Pediatric Heart Network Fontan Cross-Sectional Study

Conclusion  Management of APCs before Fontan shows marked practice variation. We did not find an association between pre-Fontan coiling of APCs and shorter postoperative hospital stay or with better late outcomes. Prospective studies of this practice are needed. (Am Heart J 2011;162:125-30.)

Background  The practice of coiling aortopulmonary collaterals (APCs) before Fontan completion is controversial, and published data are limited. We sought to compare outcomes in subjects with and without pre-Fontan coil embolization of APCs using the Pediatric Heart Network Fontan Cross-Sectional Study database which enrolled survivors of prior Fontan palliation.

Methods  We compared hospital length of stay after Fontan in 80 subjects who underwent APC coiling with 459 subjects who did not. Secondary outcomes included post-Fontan complications and assessment of health status and ventricular performance at cross-sectional evaluation (mean 8.6 ± 3.4 years after Fontan).

Results  Centers varied markedly in frequency of pre-Fontan APC coiling (range 0%-30% of subjects, P < .001). The coil group was older at Fontan (P = .004) and more likely to have single right ventricular morphology (P = .054) and pre-Fontan atrioventricular valve regurgitation (P = .03). The coil group underwent Fontan surgery more recently (P < .001), was more likely to have a prior superior cavopulmonary anastomosis (P < .001), and more likely to undergo extracardiac Fontan connection (P < .001) and surgical fenestration (P < .001). In multivariable analyses, APC coiling was not associated with length of stay (hazard ratio for remaining in-hospital 0.91, 95% CI 0.70-1.18, P = .48) or postoperative complications, except more post-Fontan catheter interventions (hazard ratio 1.74, 95% CI 1.04-2.91, P = .03), primarily additional APC coils. The groups had similar outcomes at cross-sectional evaluation.

Conclusion  Management of APCs before Fontan shows marked practice variation. We did not find an association between pre-Fontan coiling of APCs and shorter postoperative hospital stay or with better late outcomes. Prospective studies of this practice are needed. (Am Heart J 2011;162:125-30.)
Retrospective studies unlikely to address Confounding Risks

• Other predictors of Fontan outcome
  – Age, pulmonary artery pressure and resistance, ventricular function, atrioventricular valve regurgitation, type of cavopulmonary connection, fenestration...

• Some of these same risk factors likely related to collateral burden
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Quantification is key

• Angiography, TD, pulmonary venous return as measurement tools
  – not precise enough or
  – Too cumbersome and therefore impractical

• What’s needed = A reliable method of quantifying collateral flow.

• Importance of collateral flow difficult to assess without a reliable way to measure it.
Quantifying SPC flow by MRI*

\[ Q_{\text{coll_syst}} = Q_{Ao} - (Q_{SVC} + Q_{IVC}) \]

\[ Q_{\text{coll_pulm}} = (Q_{RPV} + Q_{LPV}) - (Q_{RPA} + Q_{LPA}) \]

\[ Q_{\text{coll}} = \frac{(Q_{\text{coll_syst}} + Q_{\text{coll_pulm}})}{2} \]

*Whitehead et al. Circ Cardiovasc Imaging, 2009
*Grosse-Wortman et al. Circ Cardiovasc Imaging, 2009
Questions

- What happens to collaterals in patients over time? ✔
- Are they bad? ✔
- What factors are associated with increased collateral development? ✔
- Does embolization work? ✔
- If embolization works, does it have a positive impact on important outcomes? ✔
What should we do with SPCs?
Let’s investigate....

• What happens to collaterals in patients over time?
  – At the time of Fontan collateral flow may become more significant
  – After Fontan SPCs decrease over several years in the population as a whole

• Are they bad?
  – Almost certainly but we need to learn much more

• What factors are associated with increased collateral development?
  – Stay tuned

• Does embolization work?
  – Probably only for a short time.

• If embolization works, does it have a positive impact on important outcomes?????
Conclusions: AP collaterals

• Now that we can reliably measure collateral flow, we are beginning to unravel the complex story of collaterals in single ventricle heart disease.

• In the near future we should be able to answer some of the important questions about current practice, ie the role of embolization therapy.
Conclusions: AP collaterals

- Prospective trials will be required to determine whether intervening on collateral vessels is a durable procedure and whether it affects short and long-term outcome.
**Acknowledgement**

*Jonathan J Rome MD  
**Kevin K Whitehead MD PhD  
**Andrew C. Glatz MD  
Yoav Dori MD PhD  
Mark A. Fogel MD  
Matthew A. Harris MD  
Marc S. Keller MD
• **Diagnostic catheterization** Pre-Glenn (stage 2) and Pre-Fontan is (still) an important tool along the “Fontan continuum”

• The trend is towards less invasive modalities (MRI)

• Combining MRI and catheterization (XMR) has the potential to answer the question regarding significance of collateral flow
Thank You
MRI measurement of Collateral Flow: Validation

(1) \[ Q_{\text{coll-syst}} = Q_{\text{Aorta}} - (Q_{\text{SVC}} + Q_{\text{IVC}}) \]

(2) \[ Q_{\text{coll-pulm}} = (Q_{\text{RPV}} - Q_{\text{RPA}}) + (Q_{\text{LPV}} - Q_{\text{LPA}}) \]

Whitehead et al. Quantification of Systemic-to-Pulmonary Collateral Flow

(Circ Cardiovasc Imaging. 2009;2:405-411.)