



Thromboembolic Disease after Fontan

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No Disclosures

Learning Objectives

- Discuss the pathophysiology of thromboembolic complications (TEC) after Fontan operation
- Discuss the prevalence, clinical presentation, and diagnosis of TEC
- Review prognostic implications, risk stratification and prophylaxis of TEC

TEC: Classification

Venous

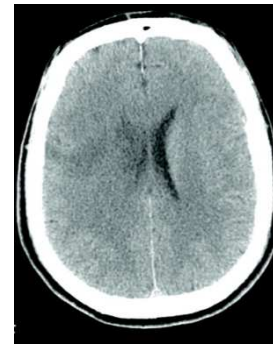
Pulmonary
embolism



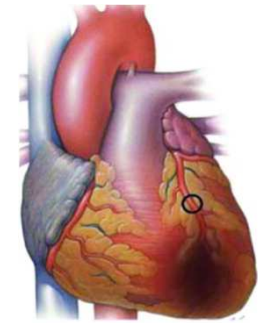
RA/Conduit
thrombus



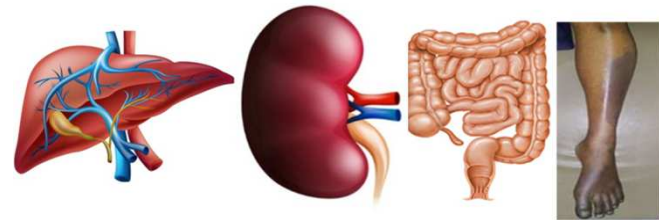
Arterial



Stroke

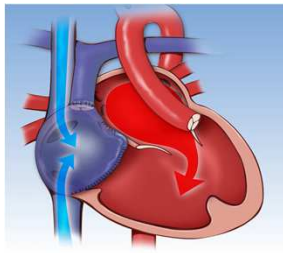


Coronary
embolus

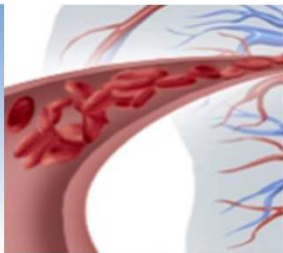


Visceral/limb infarct

TEC: Pathophysiology



'Sluggish' blood flow



Erythrocytosis

TEC

Fenestration/shunt



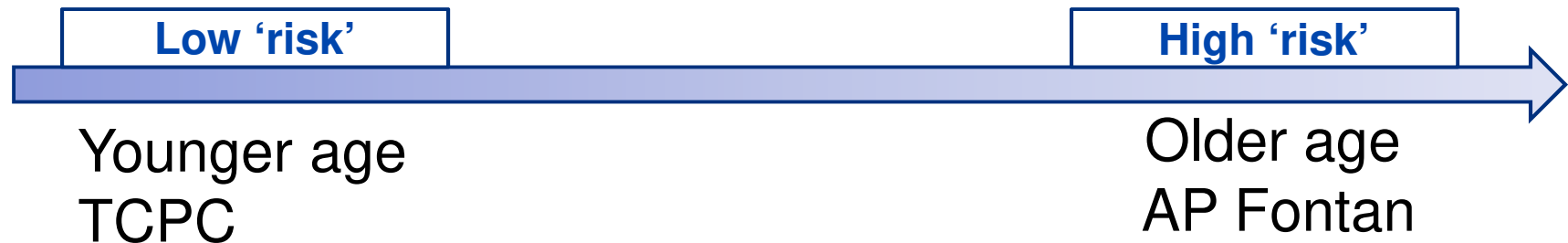
Hereditary clotting disorders



TEC: Prevalence

Prevalence: 5 – 33%

- Definition of TEC
- Method of diagnosis
- Age
- Type of Fontan



TEC: Prevalence

Low 'risk'



- Younger age
- TCPC

- N=592
- Age 4y/FU 5y
- TCPC 87%
- Echo diagnosis

9%

- N=570
- Age 5y/FU 7y
- TCPC 100%
- Multimodality

4%

- N=500
- Age 4y/FU 4y
- TCPC 100%
- Multimodality

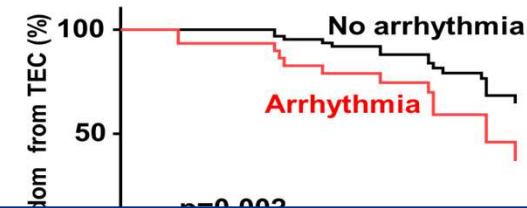
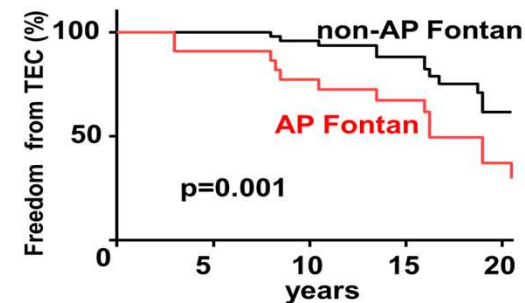
1%

TEC: Prevalence

High 'risk'

- N=387 adults
- Current age 28y
- Fontan 14y
- AP Fontan 73%

25%



33%

Balling, JTCVS 2000

16%

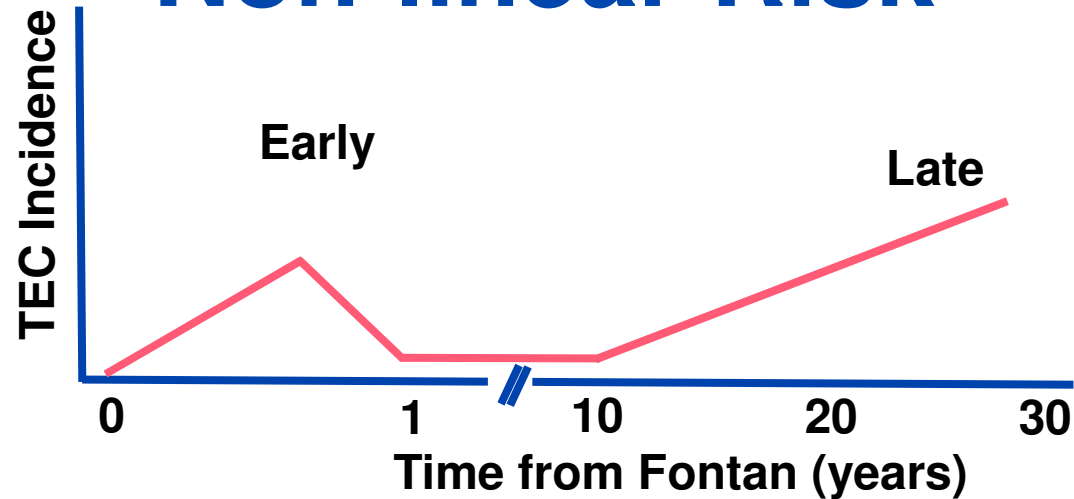
Tsang, Cardiol 2001

17%

Grewal, CHD 2013

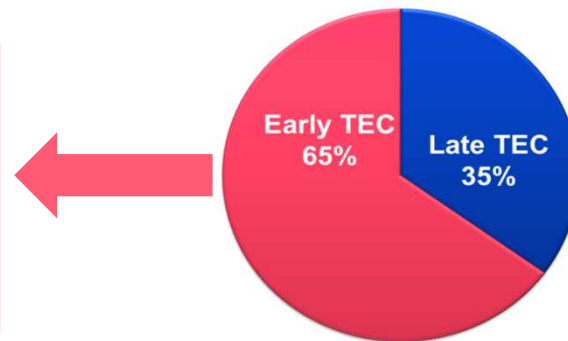
Egbe et al, Am Heart J 2017

Non-linear Risk



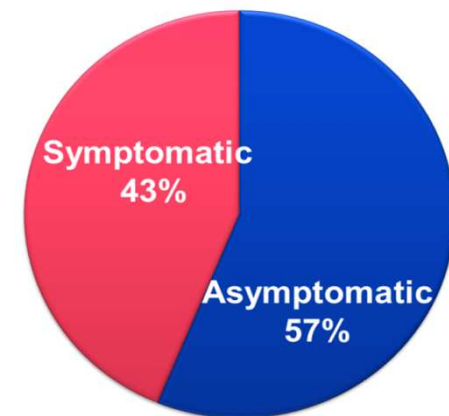
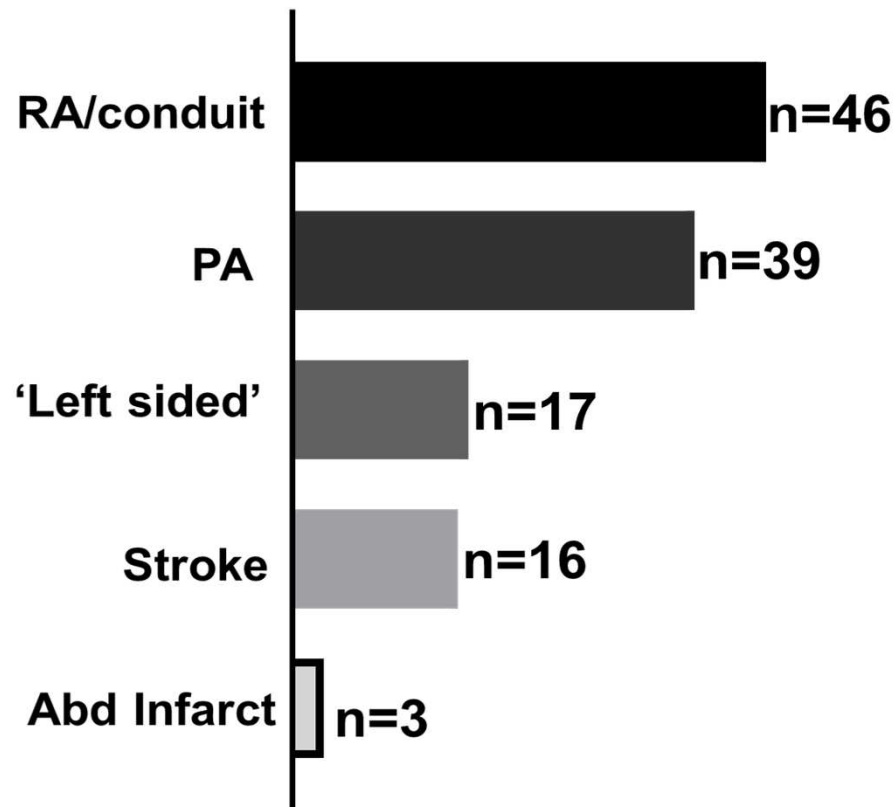
Early TEC

- Periop factors
 - Hemodynamics
 - ECMO
- Independent of Fontan type



- N=592
- TEC=52 (9%)

Clinical Presentation-Late TEC

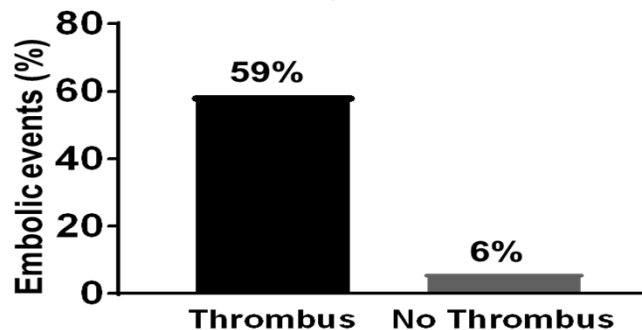


Thrombus vs Embolism

Thrombus N=46	Systemic venous atrium Fontan conduit
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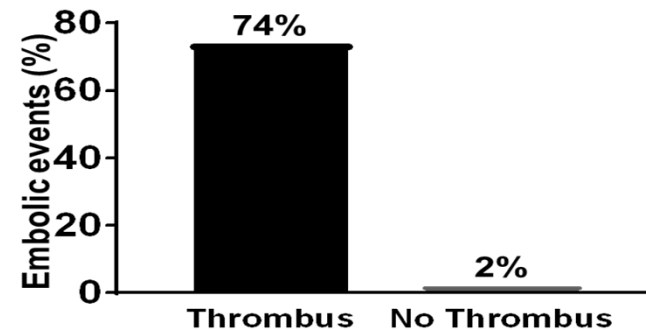
Embolism N=39	Pulm Embolism
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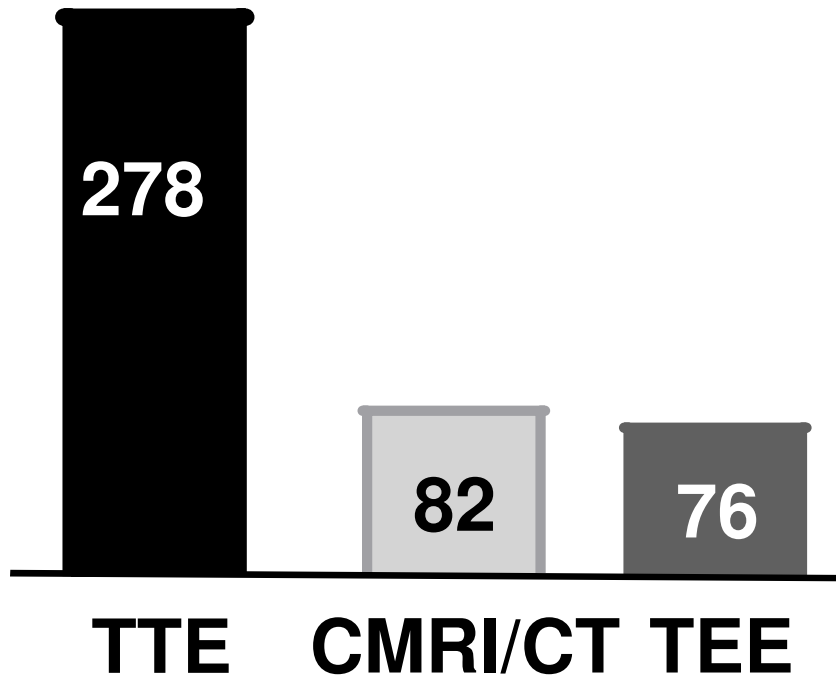
Thrombus N=17	Pulm venous atrium/ ventricle ('left sided')
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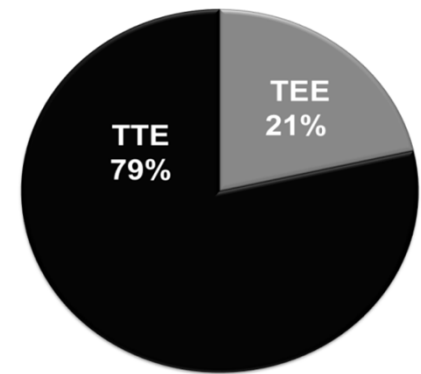
Embolism N=19	Stroke Visceral/limb infarct
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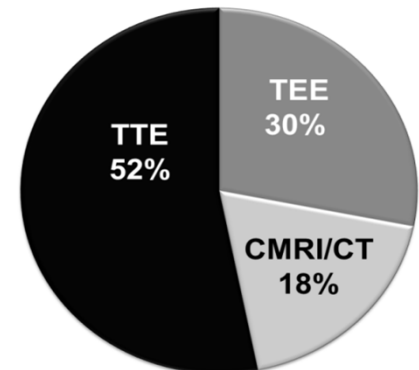
TEC Diagnosis



- Pulm venous atrium/ventricle



- Systemic venous atrium
- Fontan conduit



Learning Objectives

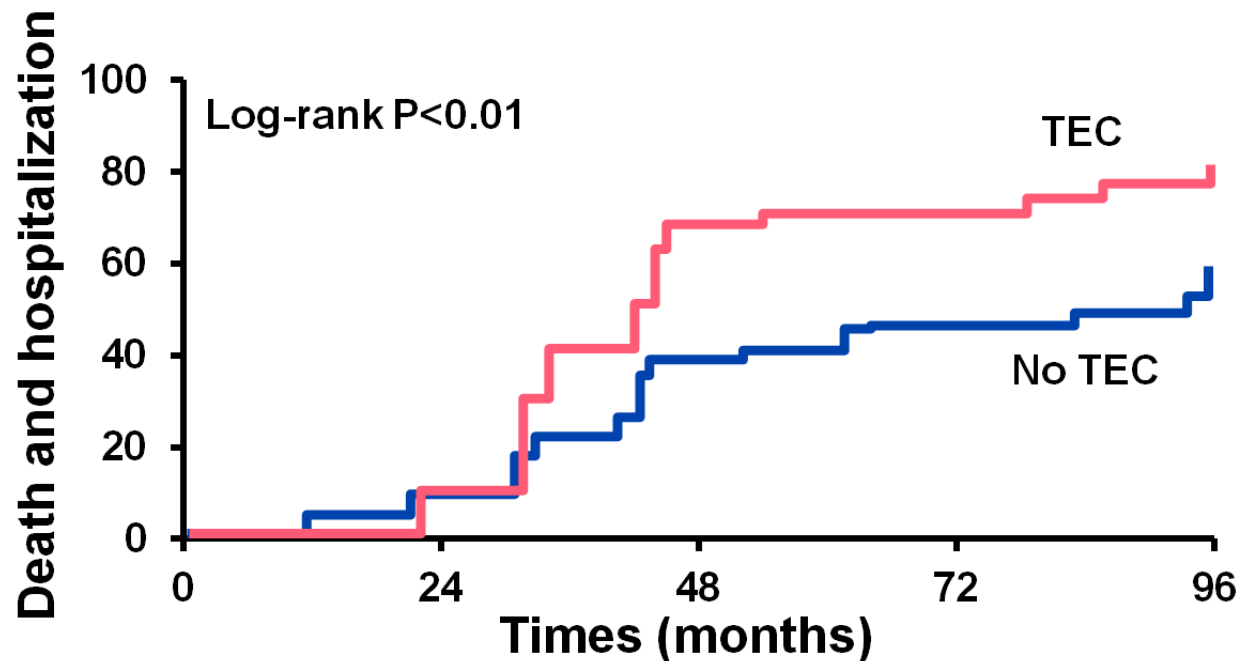
- Discuss the pathophysiology of thromboembolic complications (TEC) after Fontan operation
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To be, or not to be



TEC: Clinical Implications

Is TEC important?



TEC Prophylaxis

Aspirin

vs

Warfarin

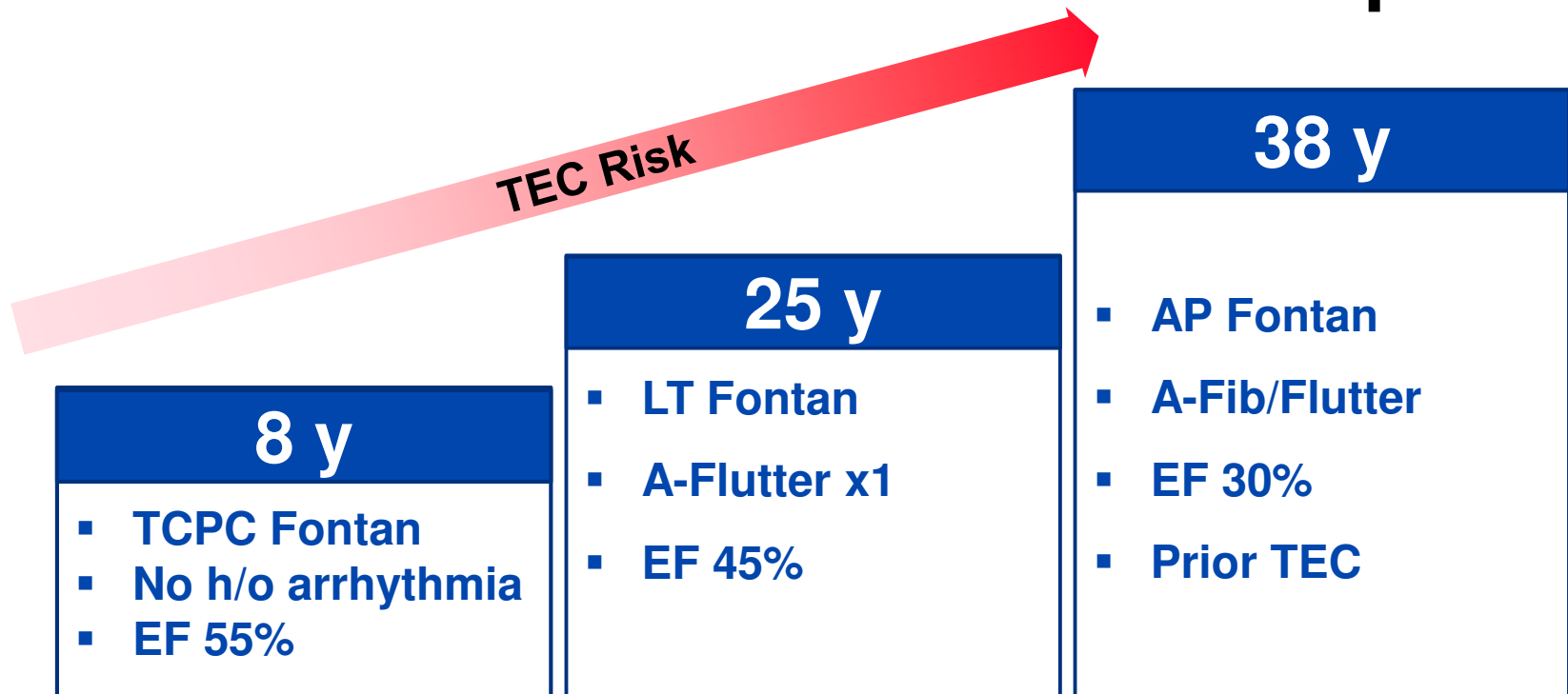
CLOTTING RISK



BLEEDING RISK

What is the risk of TEC in a Fontan?

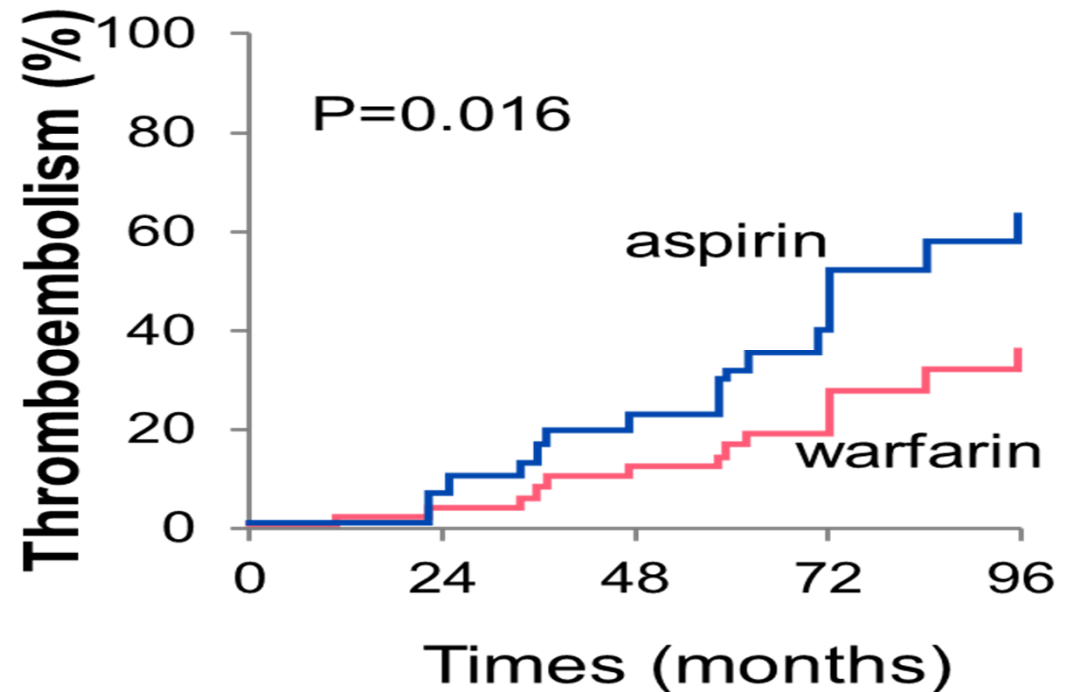
All Fontans are not created equal!



TEC Prophylaxis

Fontan with arrhythmia

- N=278
- Age: 31 ± 9
- AP Fontan 81%



Direct Oral Anticoagulants (DOACs)

NOTE Registry

- Prospective Registry for DOAC use in ACHD pt
- 33 sites in North America and Europe

NOTE Registry-Fontan Arm

70 adult patients

- APC 23 (33%)
- TCPC 47 (67%)



Outcomes

- TEC 2.1%/y
- Major bleeding 1%/y

Mayo Cohort (Warfarin)

- TEC 2.9%/y
- Major bleeding 0.6%/y

Conflicting Data

Pediatric
Cardiology

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,†

Thrombotic
Procedures
Appropriate

Ralf G.

Peter Hanrahan, MD

Eberhard G. Müh

No difference between aspirin and warfarin after extracardiac Fontan in a propensity score analysis of 475 patients

Ajay J. Iyengar^{a,b,c}, David S. Winlaw^{d,e}, John C. Galati^{b,f}, Gavin R. Wheaton^g, Thomas L. Gentles^h,
Leeanne E. Griggⁱ, Robert N. Justo^j, Dorothy J. Radford^k, Chantal Attard^l, Robert G. Weintraub^{b,c,m},
Andrew Bullockⁿ, Gary S. Sholler^{d,e}, David S. Celermajor^{e,o} and Yves d'Udekem^{a,b,c,*}
on behalf of the Australia and New Zealand Fontan Registry

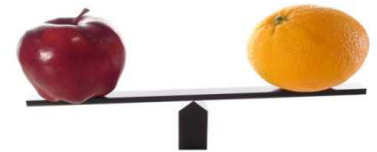
Long-term

Thrombotic and Embolic Complications Associated With Atrial Arrhythmia After Fontan Operation

Role of Prophylactic Therapy

Alexander C. Egbe, MD, MPH,^a Heidi M. Connolly, MD,^a Christopher J. McLeod, MB BCh, PhD,^a

Fontans are not created Equal



Future Directions

Fontan 'Phenotyping'

- Time (age, duration of Fontan)
- Fontan connection
- Comorbidities (FAD)
- Standardized definition of TEC
- Hemodynamics



Risk score

TEC Risk Score

CHA₂DS₂-VASc Score for Atrial Fibrillation Stroke Risk ☆

Calculates stroke risk for patients with atrial fibrillation, possibly better than the [CHADS₂ score](#).

When to Use ▼ Pearls/Pitfalls ▼ Why Use ▼

Age: <65 0, 65-74 +1, ≥75 +2

Sex: Female +1, Male 0

CHF history: No 0, Yes +1

Hypertension history: No 0, Yes +1

Stroke/TIA/Thromboembolism history: No 0, Yes +2

Vascular disease history: No 0, Yes +1

Diabetes history: No 0, Yes +1

Result:

FONTAN TEC SCORE

Atriopulmonary Fontan	No 0	Yes +1
Atrial arrhythmia	No 0	Yes +2
Fenestration	No 0	Yes +2
Ejection fraction <30%	No 0	Yes +1
Spontaneous echo contrast	No 0	Yes +1
Prior thromboembolism	No 0	Yes +2

Score ≥2: TEC ≥7.3% per year



Take Home Points

At risk for TEC

- Hemodynamics
- Arrhythmia
- Liver Disease
- Genetics
- Cyanosis/fenestration

Clinical Presentation

- Venous > Arterial
- Asymptomatic presentation (52%)

Clinical Implications

- ↑ Risk of Death/Hospitalization

Accurate diagnosis

- You may need more than TTE

Prophylaxis & Rx

Warfarin is safe & effective

- Late TEC/High risk group

DOAC:? insufficient data

RISK SCORE IS CRITICAL



Questions & Discussion

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