Adult Congenital Heart Disease; What Every Practitioner Should Know

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% Echos by Age
Children’s Hospital, Boston

S Colan: Personal Communication

<1y ➔ 18y
Birth Prevalence of CHD Subtypes Over Time

Worldwide, birth prevalence of CHD exceeds 9 per 1000 live births. In North America, prevalence rate approximates 7 per 1000 live births (40,000 graduates / year to ACHD care)
Prevalence of Congenital Heart Disease in the General Adult Population: the EXPLOSION

CHD prevalence 12/1000 children   CHD prevalence 6.07 / 1000 adults

20,000 n€  55-140,000 new pts/yr
5% in  9% increase/yr


Marelli, et al. Circ 2014;130:749-756
Adult Congenital Heart Disease: Trends in Hospitalization

88%↑ / 33%↑: ACHD/PCHD
112%↑ / 53%↑: simple/complex

O’Leary J et al. JAMA 2013
Is ACHD becoming “AHD”?

![Age at death (years) distribution for two different years: 1987-1988 and 2004-2005. The graph shows the proportion of all deaths across different age groups.]
ACHD: A Population “In Need”

2-3x ↑ hospitalizations

Hospitalizations/1000 persons

Severe CHD: 354
Other CHD: 208
Quebec Population: 103

P<0.0001

GP Outpatient visits
0.91 (0.87, 0.96)

Cardiologist outpatient visits
2.24 (2.06, 2.45)

Interaction with age (p=0.0001)
Age 18-40: RR=2.39 (2.16, 2.65)
Age 41-64: RR=1.67 (1.38, 2.01)
Age 65+: RR=1.30, (0.92, 1.85)

Specialist* outpatient visits
1.06 (1.00, 1.12)

ED visits
1.09 (1.03, 1.17)

Hospitalizations days
1.30, (1.19, 1.43)

Days in critical care
2.12 (1.80, 2.50)

GP indicates General Practitioner; RR, Rate Ratio; ED, Emergency Department; CI indicates confidence interval

ACHD Population

complex

moderate
As we develop improved metrics of R-heart function (CMR, CPET, serologies), we may learn how to best preserve outcomes and when to intervene (PVR / TVR / repeat conduit for TOF, TGA, Ebstein; targeted PAH Rx for ‘PHT’).
As we develop improved metrics of R-heart function (CMR, CPET, serologies), we may learn how to best preserve outcomes and when to intervene (PVR / TVR / repeat conduit for TOF, TGA, Ebstein; targeted PAH Rx for ‘PHT’)

CURRENT PRESENTATION:
40 y/o F w/ congenital rubella syndrome and TOF s/p repair, now with severe PR and RV dilation for PVR

ANATOMY:
(S,D,S) TOF/PS, LAA w/ NB

SIGNIFICANT CARDIAC HISTORY:
SURG (4 y/o, Michigan): TOF repair w/ VSD patch and RVOT patch (no report available)
CATH & EPS (9/20/12, DP/DM): see current data

RECENT STATUS/OTHER HISTORY:
Clinically well overall, but with limited exercise tolerance (1 flight of stairs). Occasional palpitations w/ anxiety, none otherwise. No CP or syncope.

Weight: 79 kg
PMHx: congenital rubella, bilateral sensorineural hearing loss, legal blindness, CP, strabismus, mlt HTN
PShx: ear and strabismus surgery
Meds: Lasix, lisinopril, celexa, allegro, fosamax, trazodone
Allergies: NKDA

CURRENT DATA:

EPS: No AP or inducible SVT. Possible dual AVN physiology. Negative atrial stim. Positive ventricular stim w/ inducible polymorphic VT.

LS (9/19/12): 25% left, 75% right

MRI (7/11/12): Unobstructed dilated RVOT. Severely pulsatile and dilated main and branch PAs. The branches extending into the lung periphery are asymmetric and diminished on the left associated with severe regurgitation in the LPA with little if any net antegrade flow. Differential pulmonary blood flow is nearly 100% to the right. Severe PR (66%). Mild TR. No significant RV HTN based on systolic septal configuration. Severely dilated RV (EDV 206 ml/m2; z 9.5) with moderately depressed systolic function (EF 32%). No significant MR or AR. Normal LV size with mildly depressed systolic function (EF 47%). Normal caliber aorta. Unobstructed LAA w/ NB. No significant APCs. No residual ASD or VSD. Normal pulmonary and systemic venous connection. Narrowing of the left main stem bronchus related to compression by the severely dilated and pulsatile proximal LPA.

PLAN:
OR 10/12
MRI

Preop
## Pulmonary Valve Replacement
### MRI Data

**Pre Op**
- $\text{RVEDV} = 206 \text{ ml/m}^2; Z = 9.5$
- $\text{RVESV} = 140 \text{ ml/m}^2; \text{EF} = 32\%$
- $\text{LVEDV} = 61 \text{ ml/m}^2; Z = -1.5$
- $\text{LVESV} = 33 \text{ ml/m}^2; \text{EF} = 47\%$
- $\text{RV M/V gm/ml} = 0.14$
- $\text{RPA Flow l/m}^2 = 2.16$
- $\text{LPA Flow l/m}^2 = 0.18$

**Post Op**
- $\text{RVEDV} = 113 \text{ ml/m}^2; Z = 2.7$
- $\text{RVESV} = 73 \text{ ml/m}^2; \text{EF} = 36\%$
- $\text{LVEDV} = 71 \text{ ml/m}^2; Z = -0.61$
- $\text{LVESV} = 31 \text{ ml/m}^2; \text{EF} = 57\%$
- $\text{RV M/V gm/ml} = 0.2$
- $\text{RPA Flow l/m}^2 = 1.8$
- $\text{LPA FLOW l/m}^2 = 0.75$
Pulmonary Valve Replacement
MRI Data

**Pre Op**
- RVEDV 206 ml/m²; Z=9.5
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- LPA Flow l/m² .18

**Post Op**
- RVEDV 113 ml/m² Z=2.7
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## Pulmonary Valve Replacement

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The Heart Late After TOF Repair: Myopathy

Results of years of severe chronic volume load and, in some, pressure load

- Right ventricle:
  - Dilatation
  - Dysfunction
  - Hypertrophy
  - Scar tissue
  - Diffuse fibrosis
- LV dysfunction
Natural History of Repaired TOF

Accelerated rates of mortality and morbidity beginning during the 3rd decade of life

Survival

Arrhythmias

Nollert et al. *JACC* 1997;30:1374

Khairy et al. *Circulation* 2010;122:868
Role of CMR in TOF

- None of the prior studies quantified ventricular volumes, function, mass, or amount of PR

- 2000s: several single-center studies have reported CMR parameters associated with poor outcome
## Prior Studies of Adverse Outcomes

### Studies with RV Measurements

<table>
<thead>
<tr>
<th>1st author</th>
<th>Reference</th>
<th>N</th>
<th>Outcomes</th>
<th>Predictors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Geva (Boston)</td>
<td><em>JACC 2004</em></td>
<td>100</td>
<td>NYHA Class</td>
<td>≥moderate RV and LV systolic function</td>
</tr>
<tr>
<td>Babu-Narayan (London)</td>
<td><em>Circulation 2006</em></td>
<td>92</td>
<td>Exercise capacity</td>
<td>RV and LV LGE</td>
</tr>
<tr>
<td>Knauth (Boston)</td>
<td><em>Heart 2008</em></td>
<td>88</td>
<td>NYHA Class, Death, VT</td>
<td>Severe RV dilation, LV or RV dysfunction</td>
</tr>
<tr>
<td>Wald (Boston)</td>
<td><em>Circulation 2009</em></td>
<td>62</td>
<td>Exercise capacity</td>
<td>Regional wall motion abnormalities</td>
</tr>
</tbody>
</table>
Who is at Risk for Major Adverse Clinical Outcomes?

<table>
<thead>
<tr>
<th>Odds Ratio (95% CI)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV EDVz ≥7</td>
<td>4.55 (1.10, 18.8)</td>
</tr>
<tr>
<td>LV EF &lt;55%</td>
<td>8.05 (2.14, 30.2)</td>
</tr>
<tr>
<td>or RV EF &lt;45%</td>
<td>5.60 (1.47, 21.2)</td>
</tr>
</tbody>
</table>

• QRS duration not an independent predictor

Knauth et al. *Heart* 2008
Overall goal: to create a large cohort that allows robust analyses to address clinically important questions in patients with repaired TOF

Multicenter cohort:

- Boston: Anne Marie Valente and Tal Geva
- Toronto: Rachel Wald
- London: Sonya Babu-Narayan
- Amsterdam: Barbara Mulder
Specific aim: to identify predictors of major adverse outcomes in a large contemporary cohort of late survivors of TOF repair

- Clinical, ECG, exercise, and MRI data
- Core CMR laboratory (CHB)
- Statistical core: Kimberlee Gauvreau
- Current analysis: 873 patients
INDICATOR: Results

- 873 patients with repaired TOF
  - 83% TOF/PS
  - 15% TOF/PA
  - 2% TOF/AVC
- Median age at repair 2.9 years
- Prior palliative shunt: 36%
- Surgical repairs:
  - Transannular patch: 52%
  - RV-PA conduit: 17%
  - Other: 31%
- Median age at last CMR 24.4 years
**INDICATOR: Results**

- 873 subjects (median follow-up since CMR 4.1 yrs)
- 32 subjects experienced a primary outcome during follow-up period
  - 28 deaths
  - 4 sustained VT
**Do CMR parameters improve risk stratification late after TOF repair beyond QRS duration?**

*Multivariable Analysis*

<table>
<thead>
<tr>
<th>Model</th>
<th>HR</th>
<th>95% CI</th>
<th>P Value</th>
<th>C Statistic</th>
<th>$R^2$</th>
</tr>
</thead>
<tbody>
<tr>
<td>QRS duration $\geq 180$</td>
<td>3.17</td>
<td>(1.42, 7.10)</td>
<td>0.005</td>
<td>0.586</td>
<td>0.03</td>
</tr>
<tr>
<td>QRS duration $\geq 180$</td>
<td>2.78</td>
<td>(1.20, 6.42)</td>
<td>0.02</td>
<td>0.853</td>
<td>0.24</td>
</tr>
<tr>
<td>RV mass/volume $\geq 0.3$</td>
<td>6.28</td>
<td>(2.89, 13.6)</td>
<td>&lt;0.001</td>
<td></td>
<td></td>
</tr>
<tr>
<td>LV EF z-score $&lt; -2$</td>
<td>3.46</td>
<td>(1.64, 7.30)</td>
<td>0.001</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

C statistic: Examines the power of a risk factor to predict an outcome
Nagelkerke $R^2$: Proportion of variability in model explained by variables
Comparison of QRS duration with CMR measurements

- QRS duration alone is a modest predictor of death or sustained VT ($C = 0.586; \ R^2 = 3.1\%$)
- CMR-measured RV parameters (RV mass/volume ratio and LV EF) substantially improve outcome prediction ($C = 0.853; \ R^2 = 24\%$)
Multivariable Analysis of Risk Factors for Death and Sustained VT (n= 33)

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<th>P Value</th>
<th>C Statistic</th>
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</thead>
<tbody>
<tr>
<td>RV mass $\geq 0.3$ g/mL</td>
<td>5.04</td>
<td>(2.30, 11.0)</td>
<td>$&lt;0.001$</td>
<td>0.832</td>
</tr>
<tr>
<td>LV EF z-score $&lt;-2$</td>
<td>3.34</td>
<td>(1.59, 7.01)</td>
<td>0.001</td>
<td></td>
</tr>
<tr>
<td>H/O atrial arrhythmia</td>
<td>3.65</td>
<td>(1.75, 7.62)</td>
<td>0.001</td>
<td></td>
</tr>
<tr>
<td>RV mass $\geq 0.3$ g/mL</td>
<td>4.17</td>
<td>(1.96, 8.86)</td>
<td>$&lt;0.001$</td>
<td>0.781</td>
</tr>
<tr>
<td>RV EF z-score $&lt;-2$</td>
<td>2.59</td>
<td>(1.18, 5.64)</td>
<td>0.02</td>
<td></td>
</tr>
<tr>
<td>H/O atrial arrhythmia</td>
<td>3.61</td>
<td>(1.77, 7.34)</td>
<td>$&lt;0.001$</td>
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C statistic: Examines the power of a risk factor to predict an outcome
Freedom from Outcome

Years After CMR

No. at Risk:
No risk factors 435 249 113 22 2 1
RV mass/vol. ≥ 0.3 96 69 43 29 13 7
RV mass/vol. ≥ 0.3 + LVEF z < -2 26 16 11 7 6 3
All three factors 15 6 5 2 1 1

P < 0.001 by log rank test
Is RV Pressure an Independent Risk Factor for Major Outcomes?

<table>
<thead>
<tr>
<th>No major adverse clinical outcome (N = 293)</th>
<th>Major adverse clinical outcome (N = 22)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV systolic pressure mm Hg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>40±18</td>
<td>64 ± 27</td>
<td>&lt;0.001</td>
</tr>
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</table>

Multivariable Cox regression model

<table>
<thead>
<tr>
<th>Variable</th>
<th>HR</th>
<th>95% CI</th>
<th>P</th>
<th>C Statistic</th>
<th>R²</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV mass z-score</td>
<td>↑0.1</td>
<td>1.08</td>
<td>(1.02, 1.14)</td>
<td>0.008</td>
<td>0.864</td>
</tr>
<tr>
<td>RV pressure</td>
<td>↑10</td>
<td>1.39</td>
<td>(1.19, 1.62)</td>
<td>&lt;0.001</td>
<td></td>
</tr>
<tr>
<td>LV EF</td>
<td>↓1.0</td>
<td>1.52</td>
<td>(1.19, 1.94)</td>
<td>0.001</td>
<td></td>
</tr>
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What Have We Learnt After INDICATOR

• Given the prognostic implication of RV hypertrophy in patients with repaired TOF:
  ‣ What are the histopathologic characteristics of myocardial remodeling in this population?
  ‣ What are relationships between hemodynamic load and RV remodeling at the tissue level
INDICATOR Patient

Heart failure death at age 66 years

RV M/V ratio 0.34
RV EF 27%
LV EF 42%

20% fibrosis
Myocardial Histopathology Late After TOF Repair

- Inspired by the results of INDICATOR
- Cardiac Registry at BCH (Nicola Pradegan, Stephen Sanders, et al.)
  - 8 heart specimens:
    - Age >14 years and >1 year after repair
    - RV pressure <1/2 systemic
    - Qp/Qs <1.5
- Measurements of myocyte diameter (hypertrophy) and fibrosis

Unpublished data
Myocardial Histopathology Late After TOF Repair

Hypertrophy: mean myocyte diameter

Right Ventricle

- Median cardiomyocyte diameter (μm)
- Time after repair (years)

- RV anterior free wall: \( r = 0.73 \, (p < 0.05) \)
- RV inferior free wall: \( r = 0.71 \, (p < 0.05) \)
- Normal hearts (Mean±1SD)

Left Ventricle

- Median cardiomyocyte diameter (μm)
- Time after repair (years)

- LV free wall: \( r = 0.93 \, (p < 0.001) \)
- Normal hearts (Mean±1SD)

(Time after repair)

A B C D E F
Myocardial Histopathology Late After TOF Repair

Fibrosis: % collagen

**Right Ventricle**

- Subendocardial RV anterior free wall: $r = 0.72$ ($p < 0.05$)
- Subepicardial RV anterior free wall: $r = 0.87$ ($p < 0.01$)

**Left Ventricle**

- Subendocardial LV free wall: $r = 0.95$ ($p < 0.001$)
- Subepicardial LV free wall: $r = 0.94$ ($p < 0.001$)

(Time after repair)
Indications and Timing of PVR

- Traditional approach (1970s to 2000s):
  - Symptoms (exercise intolerance, CHF, syncope)
  - High-grade ventricular ectopy (e.g., VT)
  - Severe ventricular dysfunction ± severe TR
Indications and Timing of PVR

- 25 adults s/p TOF repair with chronic PR
- Severe RV dilatation and dysfunction
- No recovery of RV dysfunction after PVR
- Conclusion: should intervene before irreversible RV dysfunction
Indications and Timing of PVR

**Risk**
- Procedural risk
- Failure of implanted PV

**Benefit**
- Life expectancy
- Quality of life
What do we know about results of PVR?

- Meta-analysis of 22 studies, 3118 patients
- 30-day mortality: 0.87%
Indications and Timing of PVR

**Risk**
- Procedural risk
- Failure of implanted PV

**Benefit**
- Life expectancy
- Quality of life
Goals of PVR

- Prolong life

- Reduce morbidity
  - Symptom-free
  - Adequate exercise capacity
  - Freedom from major arrhythmias
What do we know about the response to PVR?

- Compared with preoperative assessment, post-PVR:
  - RV EDVi and RV ESVi decrease ~30-40%
  - RV ejection fraction does not change significantly
  - LV EDVi increases slightly; LVEF does not change
  - QRS duration: slight decrease or no change
  - Exercise capacity by EST: no change
  - Symptoms consistently improve
  - Survival? Freedom from VT?
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- Survival? Freedom from VT?
Goals of PVR

- Prolong life
- Reduce morbidity
  - Symptom-free
  - Adequate exercise capacity
- Freedom from major arrhythmias
Lessons

- Failure to intervene before irreversible electromechanical RV myopathy is associated with poor clinical outcomes, including death and sustained VT.

- Similarly, delaying PVR until severe RV dilatation and dysfunction occurs is associated with a high likelihood of inadequate RV remodeling.
Sequelae of TOF surgery are associated with late morbidity and mortality, with increasing risk after the second decade of life.

- RV hypertrophy and dysfunction, LV dysfunction, and onset of atrial tachyarrhythmias are independent risk factors for death and sustained VT.
- Delaying PVR until ventricular function deteriorates is associated with poor late outcomes.
- PVR should be performed before the RV sustains an irreversible damage.
Repaired TOF or similar physiology with ≥moderate PR (regurgitation fraction ≥25% by MRI)

Cardiac Symptoms with ≥1 of the following:

- RVEDVi >150 ml/m² or Z-score >4 or RV/LV EDV ratio ≥2
- RVESVi >80 ml/m²
- RV ejection fraction <47%
- LV ejection fraction <55%
- Large RVOT aneurysm
- QRS duration >160 ms
- Sustained tachyarrhythmia
- Other hemodynamically significant abnormalities:
  - RVOT obstruction with RV systolic pressure ≥0.7 systemic
  - Severe branch pulmonary artery stenosis (<30% flow)
  - ≥Moderate tricuspid regurgitation
  - Left-to-right shunt from residual ASD or VSD with Qp/Qs ≥1.5
  - Severe aortic regurgitation
  - Severe aortic dilatation (diameter ≥5 cm)

Asymptomatic with ≥2 of the following:

- RVEDVi >150 ml/m² or Z-score >4 or RV/LV EDV ratio ≥2
- RVESVi >80 ml/m²
- RV ejection fraction <47%
- LV ejection fraction <55%
- Large RVOT aneurysm
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As we develop improved metrics of R-heart function (CMR, CPET, serologies), we may learn how to best preserve outcomes and when to intervene (PVR / TVR / repeat conduit for TOF, TGA, Ebstein; targeted PAH Rx for ‘PHT’)

CURRENT PRESENTATION:
48 yo F w/ hx PA/IVS, Ebstein's Anomaly s/p Brock pulmonary valvotomy, Tricuspid valve repair with severe TR and moderate RV dysfunction s/p recent ablation for AF. Flutter for TV repair, possible PV replacement, R Atrial Maze

ANATOMY:
(S,D,S) Ebstein's Anomaly, i.e. IVS, ASD-2

SIGNIFICANT CARDIAC HISTORY:
SURG (10/14/1966, 3 do): Brock Pulmonary Valvotomy
CATH (10/14/1966, 3 do)
CATH (2/24/1969, 2 yo)
CATH (2/20/1985, 18 yo): Mod-severe TR. Regionally dec LV function

RECENT STATUS/OTHER HISTORY:
Recent Status: Increased frequency of atrial arrhythmias requiring multiple cardioversions over last year. s/p EPS w/ Ablation 1 month ago, no breakthrough arrhythmia since. Baseline fatigue. Fully saturated
SHx: Married
Weight: 74.9 kg
Meds: Bisoprolol, Rivaroxaban, Lorazepam prn
NKDA

CURRENT DATA:
CMR (5/22/2014): Ebstein's anomaly of TV, apical displacement of septal leaflet. Absent attachments of anterior and deficient post leaflet to anomalous RV muscle bundle. Valve opens toward infundibulum. No TS, Severe TR (RF = 53%). Dilated unobstructed RVOT, branch PAs. Mild PR (RF = 14%). Severe RV dilation (RV EDV = 172.7 ml/m2, Z= 7.1). Volume of functional chamber = 115.6 ml/m2. Global RV systolic dysfunction, regional dysfunction of atrialized chamber, including septum. RV EF = 37-38%. Severe RA enlargement, dilated CS and IVC. Mild MR. normal LV size, mild systolic dysfunction (LV EF = 42%) w/ prominent apical trabeculation. Mild LVP contraction between dAo and LA.
EST (10/24/2014): Below avg endurance, severely depressed peak VO2 (15.6,57% predicted), due to inability to augment SV and increase HR. PVCs, Couplet PACs, atrial tachycardia in recovery.

UPCOMING SCHEDULED STUDIES/PLAN:
SURG (6/16/2015)

Conditions: EP Study/HD Cath, GA, RA.
PreOp
Indications for Surgery

• Symptoms
Indications for Surgery

• Symptoms
  – Exercise Intolerance?
Indications for Surgery

• Symptoms
  – Exercise Intolerance?
• Arrhythmias
Indications for Surgery

• Symptoms
  – Exercise Intolerance?

• Arrhythmias
  – WPW?
  – VT?
Indications for Surgery

- Symptoms
  - Exercise Intolerance?

- Arrhythmias
  - WPW?
  - VT?

- LV Dysfunction?
Indications for Surgery

• Symptoms
  – Exercise Intolerance?

• Arrhythmias
  – WPW?
  – VT?

• LV Dysfunction?

• Degree of TR
Operative Data
Cone Operation

Operative

• CPB time 265 minutes
• Cross Clamp 190 minutes
• Fibrillatory arrest time 22 minutes
• Right Atrial Cryo MAZE
Operative Data
Cone Operation

**Operative**
- CPB time 265 minutes
- Cross Clamp 190 minutes
- Fibrillatory arrest time 22 minutes
- Right Atrial Cryo MAZE

**Post Operative**
- Junctional rhythm
  - NSR with Incomplete RBBB
- Extubated POD 1
- Transferred to floor POD 2
- Discharged POD 6
  - Bisoprolol
Ebstein’s Anomaly: Features relevant to the surgeon

- Size of atrialized portion of RV
- Extent of delamination of anterior leaflet
- Status of RVOT
- Cyanosis
- LV dysfunction
- Imaging: Echo and MRI
Surgical approach: Cone repair concept
To create a cone shaped valve, the tricuspid valve leaflets must be detached from the AV valve annulus, inferior free wall of the right ventricle, and from any tethering chords or muscles to the RV free wall, leaving only leaflet edge chords attached apically.
Cone repair
## Results

<table>
<thead>
<tr>
<th></th>
<th>Cone Procedure N = 19</th>
<th>Convention al Surgery N = 13</th>
</tr>
</thead>
<tbody>
<tr>
<td>Deaths:</td>
<td></td>
<td></td>
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<tr>
<td>- Early deaths (&lt;30 days)</td>
<td>0</td>
<td>0</td>
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<tr>
<td>- Late deaths</td>
<td>2 (11%)</td>
<td>1 (8%)</td>
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<tr>
<td>Reoperation¹</td>
<td>3 (16%)</td>
<td>1 (8%)</td>
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<tr>
<td>New onset dysrhythmias:</td>
<td></td>
<td></td>
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<tr>
<td>- anti-arrhythmic medication</td>
<td>1 (5%)²</td>
<td>1 (8%)²</td>
</tr>
<tr>
<td>- pacemaker implantation for CHB</td>
<td>2 (11%)</td>
<td>2 (15%)</td>
</tr>
<tr>
<td>Prophylactic pacemaker/AICD implantation</td>
<td>1 (5%)³</td>
<td>2 (15%)⁵</td>
</tr>
</tbody>
</table>
Late results: vena contracta

Cone Group – vena contracta

Conventional Group – vena contracta
Thank You