ANESTHESIA FOR CONGENITAL HEART DISEASE PATIENTS IN NON-CARDIAC SURGERY

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2018 Nov 26 @UCSF Mission Bay, Residents/Fellows’ Lecture
CONGENITAL HEART DISEASE SCOPE

- 1:125 live births. 90% live unto adulthood
- Increased perioperative morbidity, cardiac arrest, and mortality
- UCSF Mission Bay 2017
  - ~170 non-cardiac surgeries in CHD patients per annum
  - ~420 cardiac Catheterizations p.a.
  - ~450 cardiothoracic surgeries p.a.
PHYSIOLOGIC CATEGORIES OF CHD

• Simple Left to Right Shunt - ASD, VSD, CAVC, PDA, AP window
  Increased Pulmonary Blood Flow
• Simple Right to Left Shunt - TOF, PA, TA, Ebstein’s
  Decreased Pulmonary Blood Flow
• Complex Shunts - TGA, Truncus, TAPVR, DORV, HLHS
• Obstructive Lesions - AS, PS, Coarct, Interrupted Arch
MIXED CIRCULATION AND SINGLE VENTRICLE CIRCULATION

Normal or series circulation
- Right atrium → Right ventricle → Lungs → Aorta
- Left atrium → Left ventricle

Parallel or balanced circulation (e.g. truncus arteriosus)
- Blood flow to lungs/body depends on balance of PVR/SVR
- Right and left atrium → Right and left ventricle → Aorta

Single ventricle circulation (e.g. total cavopulmonary circulation)
- SVC → Lungs → Pulmonary artery → Common atrium → Common ventricle → Aorta
- IVC, inferior vena cava; SVC, superior vena cava.

PVR, pulmonary vascular resistance; SVR, systemic vascular resistance.

ANAESTHESIA AND INTENSIVE CARE MEDICINE 16:8
Savini Wijesingha
Michelle White
PHYSIOLOGIC STATUS OF CHD

- Cardiac Function - Severe: 10% perioperative CA, 96% inotropes
- Arrhythmia - Ventricular ectopy + SV 30% sudden death
- Pulmonary HT - PAP >25mmHg at rest, >30mmHg exercise
- Cyanosis - polycythemia, hyperviscosity,
- Venous Capacitance (↓) - Stress Volume (↑) in Fontan
HOW TO STRATIFY?

• Complexity of Lesion
  • Balanced Circulation (“Mixed”)
  • Single Ventricle
  • Cardiomyopathy (Severe Cardiac Failure)
  • Aortic Stenosis
  • Arrhythmia
• ASA Physical Status and Cardiac Function Status
• Age, Type of Surgery, LOS prior to Surgery,
## Risk classification of children with heart disease undergoing non-cardiac surgery

<table>
<thead>
<tr>
<th>High risk</th>
<th>Intermediate risk</th>
<th>Low risk</th>
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<tbody>
<tr>
<td>Complex lesions</td>
<td>Major surgery</td>
<td>Simple lesions</td>
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<tr>
<td>Physiologically poorly compensated and/or</td>
<td>Under 2 years old</td>
<td>Physiologically normal or well compensated</td>
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<td>presence of major complications</td>
<td>Emergency surgery</td>
<td>Minor (or body surface) surgery</td>
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<td>Preoperative hospital stay more than 10—14 days</td>
<td>Over 2 years old</td>
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<td>ASA physical status IV or V</td>
<td>Elective surgery</td>
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<tr>
<td></td>
<td></td>
<td>Preoperative hospital stay less than 10 days</td>
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<tr>
<td></td>
<td></td>
<td>ASA physical status I — III</td>
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<tr>
<td>Conduction Abnormalities</td>
<td>Low risk</td>
<td>Moderate risk</td>
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<tr>
<td>Repaired atrial or ventricular septal defect</td>
<td>Wolff Parkinson White</td>
<td>Long QT syndrome</td>
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<td>Mild regurgitation or stenosis of a single valve</td>
<td>Simple un repaired lesions such as ventricular or atrial septal defect</td>
<td>Complex cardiac defects with full repair</td>
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<tr>
<td></td>
<td>Single ventricle with Glenn or Fontan palliation</td>
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<tr>
<td>Pulmonary hypertension</td>
<td>New York Heart Association functional class I</td>
<td>Normal cardiac index</td>
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<tr>
<td>Miscellaneous</td>
<td>Heart or lung transplant</td>
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</tbody>
</table>
• 173 GAs in 71 Complex CHD patients (RACHS-1 score 6) over 5 years

• Induction instability associated with Unrepaired or Stage 1 repair, case complexity, preop use of ACEi

• Maintenance instability associated with case complexity, preop use of digoxin or inotropes

• Postop LOS associated with preop LOS >14 days, and moderate ventricular dysfunction; digoxin was protective.
**NSQIP-P STRATIFICATION OF CHD**

Table A.2
NSQIP-P definition and classification of CHD severity.

<table>
<thead>
<tr>
<th>Classification</th>
<th>Definition and Criteria</th>
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| Minor CHD      | • Cardiac condition with or without medication and maintenance (e.g., atrial septal defect, small-to-moderate ventricular septal defect with no symptoms)  
• Repair of congenital heart defect with normal cardiovascular function and no medication |
| Major CHD      | • Repair of congenital heart defect with residual hemodynamic abnormality with or without medications (e.g., Tetralogy of Fallot with wide open pulmonary insufficiency, hypoplastic left heart syndrome including stage 1 repair) |
| Severe CHD     | • Uncorrected cyanotic heart disease  
• Patients with any documented pulmonary hypertension  
• Patients with ventricular dysfunction requiring medications  
• Listed for heart transplant |

NSQIP-P = National Surgical Quality Improvement Program Pediatrics version; CHD = congenital heart disease.
Post-Operative Outcomes in Children With and Without Congenital Heart Disease Undergoing Noncardiac Surgery

David Faraoni, MD, PhD, David Zurakowski, PhD, Daniel Vo, MD, Susan M. Goobie, MD, Koichi Yuki, MD, Morgan L. Brown, MD, PhD, James A. DiNardo, MD

- 2012 pediatric database of the American College of Surgeons National Surgical Quality Improvement Program: NSQIP – Pediatrics (n = 51008)
- 4520 CHD => 2805 CHD after propensity matching:
  - Severe 417, Major 1272, Minor 1272
  - Subgroups matched for surgical complexity
- Overall mortality higher in Severe (8.2% vs. 1.7%, p < 0.001) and Major (3.9% vs. 1.2%, p = 0.001)
- No difference between Minor CHD and Matched Controls.
NSQIP 2012-2013 (training model) & NSQIP 2014 (testing model), n=183423

Severe or Major CHD population

Primary outcome: In-Hospital Mortality

Logistic Regression = > 8 risk factors (OR, all p < 0.007)

Risk Stratification Score (AUC 0.831 [.787-0.875])

- 0 - 3 (OR 1.54 [0.78-3.04])
- 4 - 6 (OR 4.19 [2.56-6.87])
- 7 - 10 (OR 22.2 [15.1-32.6])
CONCLUSIONS: Our study demonstrates that, in addition to preoperative markers of critical illness (eg, inotropic support, mechanical ventilation, preoperative cardiopulmonary resuscitation, and acute or chronic kidney injury), the type of lesion (eg, single-ventricle physiology) and the functional severity of the heart disease (eg, severe CHD) are strong predictors of in-hospital mortality in children undergoing noncardiac surgery. (Anesth Analg 2016;123:824–30)

1) Emergency procedure, 2) previous surgery 30-day,
3) Severe CHD, 4) Single-Ventricle,
5) Inotropic support, 6) Preoperative CPR, 7) Acute or Chronic Kidney Injury, 8) Mechanical Ventilation.
Patients with single ventricle physiology undergoing noncardiac surgery are at high risk for adverse events
Morgan L. Brown, James A. DiNardo & Kirsten C. Odegard
Division of Cardiac Anesthesia, Department of Anesthesiology, Boston Children’s Hospital, Boston, MA, USA

The Frequency of Cardiac Arrests in Patients with Congenital Heart Disease Undergoing Cardiac Catheterization
Kirsten C. Odegard, MD, Lisa Bergersen, MD, MPH, Ravi Thiagarajan, MD, Laura Clark, RA, Avinash Shukla, MBBS, David Wypij, PhD, and Peter C. Laussen, MBBS
ANESTHETIC MANAGEMENT IN GENERAL

- **Plan** (steps detailed on right side box)
- Native and Current Anatomy
- SBE Prophylaxis
- “Bubble-less” injection and De-Airing
- Invasive and Other Monitoring
- Fluid, Hemodynamics
  - Cardiac Physiology and State
  - SVR, PVR, HR, Contractility, Preload => CO
  - Venous Return (S:P) => Q_S:Q_P => CO
- Postoperative Care
- Other Unique Factors

- Plan and Do
- Assessment and Optimization
- Induction
- Maintenance
- **Critical Points**
- Emergence
- Transfer
- Follow-up and Review
TOF WITH RESIDUAL DEFICIT – TRACHEAL DILATION

- E.F. 20 y.o. F
  “w/ remote history of TOF/PA s/p repair with fenestrated VSD & artificial pulmonary valve and MAPCAs s/p unifocalization”
- PHT: WHO Illb, suprasystemic RVHT
- Previous endocarditis
- Tadalafil, nocte NC oxygen.
- Aspirin, prednisone, bronchodilators, bisoprolol
- Dyspnoea on exertion
- 36.3kg, 80% RA, no PACU BPs or HR

- Tracheal Dilation and Laser Debridement
- What information would you like?
- How will you anesthetize this patient?
HLHS S/P NORWOOD FOR DLB

- 3.8kg, 2 mth s/p Stage 1, CICU.
- CHARGE; Tachyarrhythmia, failed extubations, backup VVI
- Milrinone, diuretics, fluconazole, cefepime, fentanyl, midazolam, L-thyroxine, amiodarone, flecainide
- 3.5 uncuffed ETT, Fi 0.4, 78%, 109, 62/32
- Artline, PICC
- Concerns? Reassurance?

- “The patient was paralyzed, sedated, and intubated…”
- “The Miller laryngoscope was used to expose the larynx…”
- “2.5 rigid ventilating bronchoscope to obtain a magnified view…”
- Normal trachea, mild carina granulation, mild stenosis of L mainstem bronchus without compression, normal R mainstem bronchus. No surgically addressable sites of airway obstruction.
BDG FOR DLB, EUA

- 19.5kg, 7 y.o. M, HLHS s/p Glenn Shunt
- Previous Tracheostomy, laryngomalacia, subglottic stenosis, mild OSA
- 80% RA,
- Single coronary, MPA band PG 80mmHg
- ECHO: good function
- s/f DLB, prior to eventual Fontan Repair

Concerns?

- "Parsons…, Lindholm…, Hopkins Rod Telescope…"

Where would you like to send him?
CHD: QUICK WORD ON DISCHARGE

Unanticipated hospital admission in pediatric patients with congenital heart disease undergoing ambulatory noncardiac surgical procedures

Koichi Yuki¹-² | Sophia Koutsogiannaki¹-² | Sandra Lee² | James A. DiNardo¹-²

- 1028 CHD patients for **ambulatory** procedures at 1 center (n 3010, 2008-2013)
- Demographics, echo function, physical function, CVS and respiratory adverse events, reasons for unplanned admissions
- Unplanned admissions - 2.7%
- Adverse CVS - 3.9%
- Adverse Respiratory - 1.8%
- IR procedures or Echo within 6 months associated with unplanned admissions
DORV/TGA FOR G-TUBE

- 7kg, 5mth M, DORV/TGA, VSD, ASD, small PVs, mild PS, PDA
- TEF repaired, vertebral and limb defects (VACTERL), duodenal atresia repair
- Previous laparotomies for gastric leak from G-Tube
- HFNC, diuretics, bronchodilators, J-feeds, methadone, lorazepam
- Monitoring?
- Induction?
- GA type?
- Other thoughts?
DORV/TGA FOR G-TUBE
HLHS S/P NORWOOD FOR G-TUBE

- 3.9kg, 2mth F, s/p Norwood/Sano,
- NEC, pleural effusions, reintubations.
- 0.5L Fi 0.3 NC,
- Feeding intolerance, s/f for Laparoscopic G-Tube
- Concerns? Goals?
HLHS S/P NORWOOD FOR G-TUBE

- Laparoscopic, with caution
- Sevoflurane, dexmedetomidine
- Ca$^{2+}$, Epi 2mcg, NaHCO$_3$·4mmol, post-induction
- PRBC 40ml for HCT 0.39
- Induction: 58 min
- Procedure: 120 min
- Laparoscopic, all the way
Outcomes of laparoscopic and open surgery in children with and without congenital heart disease

David I. Chu a,*,1, Jonathan M. Tan b,*, Peter Mattei c, Allan F. Simpao b,*, Andrew T. Costantino e, Aseem R. Shukla a, Joseph W. Rossano f, g, Gregory E. Tasian a, g, h

A B S T R A C T

Background: Children with congenital heart disease (CHD) often require noncardiac surgery. We compared outcomes following open and laparoscopic intraabdominal surgery among children with and without CHD.

Methods: We performed a retrospective cohort study using the 2013–2015 National Surgical Quality Improvement Project-Pediatrics. We matched 45,012 children <18 years old who underwent laparoscopic surgery to 45,012 children who underwent open surgery. We determined the associations between laparoscopic (versus open) surgery and 30-day mortality, in-hospital mortality, 30-day morbidity, and postoperative length-of-stay.

Results: Among children with minor CHD, laparoscopic surgery was associated with lower 30-day mortality (Odds Ratio [OR] 0.34 [95% Confidence Interval 0.15–0.79]), in-hospital mortality (OR 0.42 [0.22–0.81]) and 30-day morbidity (OR 0.61 [0.50–0.73]). As CHD severity increased, this benefit of laparoscopic surgery decreased for 30-day morbidity (p-trend = 0.01) and in-hospital mortality (p-trend = 0.05), but not for 30-day mortality (p-trend = 0.27). Length-of-stay was shorter for laparoscopic approaches for children at cost of higher readmissions. On subgroup analysis, laparoscopy was associated with lower odds of postoperative blood transfusion in all children.

Conclusions: Intraabdominal laparoscopic surgery compared to open surgery is associated with decreased morbidity in patients with no CHD and lower morbidity and mortality in patients with minor CHD, but not in those with more severe CHD.

Level-of-evidence: Level III: Treatment Study.
Minor CHD: Laparoscopic associated with lower 30-day mortality and 30-day morbidity.

Major or Severe CHD: No difference, viz. no real benefit of laparoscopy over open surgery.
EBSTEIN’S + APS FOR NISSEN’S

• 4.4kg, 2mth F, Severe Ebstein’s Anomaly, Pulmonary Atresia
• Aorto-Pulmonary Shunt, Modified Starnes’ Procedure (RV exclusion) = Single LV

• Concerns with Shunts?
EBSTEIN’S + APS FOR NISSEN’S: LAPAROSCOPY
EBSTEIN’S + APS FOR NISSEN’S: ECMO
EBSTEIN’S + APS FOR NISSEN’S: SHUNT DOWN

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
</tr>
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<tbody>
<tr>
<td>1114</td>
<td>Induction</td>
</tr>
<tr>
<td>1207</td>
<td>Anesthesia Ready</td>
</tr>
<tr>
<td>1240</td>
<td>Procedure(s) Start</td>
</tr>
<tr>
<td>1351</td>
<td>Quick Note</td>
</tr>
<tr>
<td></td>
<td>Sat trending down from high 80s/low 90s to low 80s... Hand ventilation started and FIO2 increased, surgeon made aware. Abdomen insufflation pressure lowered from 12mmHg to 10mmHg.</td>
</tr>
<tr>
<td>1355</td>
<td>Quick Note</td>
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<tr>
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<td>Continues to deteriorate despite hand ventilation and 100% FIO2, abdomen de-sufflated, trocars out, help called</td>
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<td>1358</td>
<td>Quick Note</td>
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<td></td>
<td>CPR started</td>
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<td>1421</td>
<td>Quick Note</td>
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<tr>
<td></td>
<td>Pacing wires attached</td>
</tr>
<tr>
<td>1431</td>
<td>Quick Note</td>
</tr>
<tr>
<td></td>
<td>ECMO Cannulation began</td>
</tr>
<tr>
<td>1457</td>
<td>Quick Note</td>
</tr>
<tr>
<td></td>
<td>On ECLS</td>
</tr>
<tr>
<td>1545</td>
<td>Procedure(s) Stop</td>
</tr>
<tr>
<td>1610</td>
<td>OUT of Room/Stop Data</td>
</tr>
<tr>
<td></td>
<td>Time patient and/or anesthesia team leave patient bedside/procedure room.</td>
</tr>
</tbody>
</table>
REPAIRED TAPVR FOR E-LAPAROTOMY

- 13 day (born 34+6), 3.45kg M
- TAPVR (vertical vein type, post-repair), emergency laparotomy
- 100%, RR 38, HR 171
- ASA5e, intubated uncuffed 3.5, inotropes epinephrine, dopamine, calcium, fentanyl

What would you do prior to OR?

- Fentanyl, vecuronium, minimal-none sevoflurane and PRBC
- Operative Findings Jejunal Perforations
REPAIRED TAPVR FOR E-LAPAROTOMY
REPAIRED TAPVR FOR CATHETERIZATION 2 WEEKS’ TIME

- 3.45kg, 4 wks M,
- ETT cuffed 3.5, inotropes similar to 2 wks’ ago, slightly increased:
- iNO 20 ppm for PHT *see echo*, dexmedetomidine, fentanyl, epinephrine, calcium, dopamine.
- s/f cardiac catheterization for increasing $O_2$ requirements and worsening agitation. Query anastomotic issues.

What would you do prior to transfer and what else do you get ready?
2018.10.4
1. s/p TAPVC repair. 2. s/p ASD patch closure. 3. s/p PDA ligation.
4. Normal left ventricular systolic function.
5. Evidence of high filling pressures and mild IVC dilation.
7. Greater than 50% systemic right ventricular pressure.
8. Normal RV systolic qualitative shortening.
9. Moderate right pleural effusion.
10. Moderate left pleural effusion.
11. Small pericardial effusion.

Arrhythmia is noted during this study.

2018.10.10
1. s/p TAPVC repair. 2. s/p ASD patch closure. 3. s/p PDA ligation.
4. Pulmonary vein stenosis of the right lower pulmonary vein, of the left upper pulmonary vein and of the left lower pulmonary vein.
5. At least systemic right ventricular pressure.
6. There is no residual atrial septal defect.
7. Mildly dilated right atrium.
8. Mild to moderate tricuspid regurgitation.
9. Moderately diminished RV systolic qualitative shortening. 10. Mild dilation RV.
11. Normal left ventricular systolic function.
12. Small pericardial effusion.
TAPVR, POST LAPAROTOMY, NOW PHT
POST CARDIAC CATH

| Intake (ml) | 140 |
| Blood Loss (ml) | 10 |
| Radiation Dose (mGy) | 33 |
| Contrast | Omnipaque 13.0ml |
| Procedure Duration | 285 min |
| Fluoro Time (minutes) | 39.0 |
| Anesthesia Type | General Anesthesia |
| DAP (mcGm²) | 113 |

**Diagnoses**
- Pulmonary vein obstruction, acquired (I28.8)
- Pulmonary vein stenosis, congenital (Q26.8)
- s/p TAPVR repair (747.41)
- Pulmonary hypertension, Secondary (I27.1)

**Indications**
4 wk old ex 34 week obstructed TAPVR to Rt SVC s/p repair (confluence anastomosis) c/b intestinal perf/surgery now with concerns or PV re-obstruction.

**Procedures**
- Cath, diagnostic right and left retrogra (93531) RFA 4
- Angiography: pulmonary artery, Pulmonary vein (93541, 93543) RFV 4

**Key Findings / Recommendations**
- suprasystemic PA pressures (150%) with marked elevated PVR (>14 WU), PA mean 60, LVED 19 mmHg, min grad across LA anastomosis, TLP, RU, LpVs. Min response to Iloprost.

**Complications:** None
THANKS AND QUESTIONS
REMEMBER TO REVIEW AND REFLECT

- **Plan**
- SBE Prophylaxis
- “Bubble-less” injection and De-Airing
- Invasive and Other Monitoring
- Fluid, Hemodynamics
  - Native-Current Anatomy and Cardiac State
  - SVR, PVR, HR, Contractility, Preload => CO
  - Venous Return (S:P) => Q_S:Q_P => CO
- Postoperative Care
- Other Critical Factors
- **Review and Reflect**
REFERENCES


• Faraoni et al. Post-Operative Outcomes in Children With and Without Congenital Heart Disease Undergoing Noncardiac Surgery. JACC. 2016;6(7): 793-801


• Yuki et al. Induction techniques for pediatric patients with congenital heart disease undergoing noncardiac procedures are influenced by cardiac functional status and residual lesion burden. J Clin Anes. 2018;50:14-17


• Brown et al. Patients with single ventricle physiology undergoing noncardiac surgery are at high risk for adverse events. Ped Anes. 2015;25:846-851