How to Make an Easy Case Hard: Managing an Adult with Fontan Physiology and Multisystem Organ Dysfunction for a Peritoneal Dialysis Catheter

Victoria Melanson MD[1], Vikas O'Reilly-Shah MD PhD[1,2]

[1]Department of Anesthesiology, Emory University; [2]Division of Pediatric Anesthesiology, Children's Healthcare of Atlanta

Introduction

Adult congenital heart disease is one of the fastest growing areas in adult cardiovascular medicine, including geriatric congenital heart disease (CHD) care.[1] Adults now outnumber children with CHD, accounting for 66% of the overall CHD population, and 60% of those with severe CHD.[1,2] In 2010, 1.4 million adults in the US were living with CHD, with 160,000 having severe disease.[3] The prevalence of severe CHD in adults has increased by 85% from the year 1985 to 2000, which suggests that improved surgical techniques and advanced medical intervention have had a positive influence on patient survival.[4]

Relevant Statistics

- 85% of CHD patients survive into adult life[5]
- In the year 2000, there were an estimated 800,000 adults with CHD in the United States
- In the next decade, an estimated 1 in 150 adults will have CHD
- VSD is the most common defect, followed by ASD and TOF

Surveys show that knowledge and comfort with adult CHD is low among anesthesiologists, even at specialized centers.[6] Knowing when to implement simple interventions like an arterial line or a TEE can help prevent adverse outcomes in this vulnerable population.[7] Becoming familiar with management of adult CHD patients may greatly influence their outcomes, both perioperatively and long term.

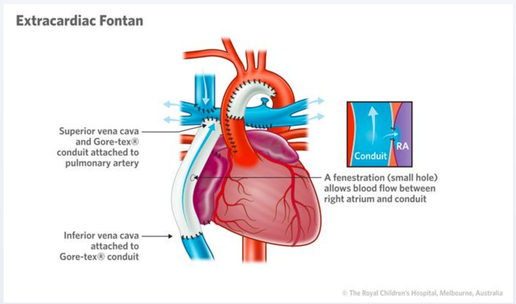
Case Report

Mr. X is a 30 year-old male weighing 64kg with a past medical history of single ventricle physiology, pulmonary hypertension, permanent pacemaker, horseshoe kidney in ESRD, and gout. He is currently under consideration for heart-liver-kidney transplant. Notably, two other referral centers did not feel he was an appropriate candidate for this transplant surgery given his complex history and anatomy. His cardiac history includes a Fontan repair (see below). Baseline SpO2 is 84% on room air. LVEF was 40% on TTE a week prior to surgery. Mr. X presents for a laparoscopic peritoneal dialysis catheter.

Notably during a recent hospitalization, Mr. X was found to have new onset atrial flutter with rapid ventricular rate, which required TEE cardioversion and transfer to the cardiac intensive care unit.

Cardiothoracic History

- Ventricular inversion / atretic pulmonary valve and VSD, s/p extracardiac Fontan (X,X,L-TGA)
- Heterotaxy syndrome with left atrial isomerism, interrupted IVC, and bilateral dual lobed lungs
- July 2005 bidirectional Glenn and extracardiac Fontan with inclusion of hepatic veins only



Intervention/Treatment

After discussion with the surgeon, the original laparoscopic surgical plan was changed to an open peritoneal dialysis catheter without the use of paralytic agents in order to ensure spontaneous ventilation. In preparation for the case, all IV access points were protected with 22-micron filters to prevent air emboli. A pre-induction arterial line was placed for hemodynamic monitoring.

The patient was induced with midazolam, low-dose ketamine for sedation without significant cardiac suppression, and sevoflurane. A magnet was placed over his pacemaker. An LMA was inserted at adequate depth of anesthesia, maintaining spontaneous ventilation. Pressure support ventilation was titrated as needed to maintain tidal volumes. Metronidazole was administered for prophylaxis prior to surgical incision.

Pain was managed via subcutaneous injection of local anesthetic per the surgical team, as well as small remifentanil boluses as needed in anticipation of surgical stimulus. Close communication between the surgical and anesthesia teams was emphasized during the case. Anesthesia was maintained with sevoflurane, and a dexmedetomidine infusion was used for opioid reduction and emergence. SpO2 ranged in the 80s to low 90s. Norepinephrine and vasopressin were the pressors of choice, and the patient required an infusion of norepinephrine throughout most of the case. From induction to emergence, the case lasted 2.5 hours.

Results

Mr. X emerged rapidly and uneventfully, and he had an uneventful PACU course. The patient was transferred to the cardiac intensive care unit for further management. The congenital heart service as well as nephrology were consulted.

The patient underwent diuresis during his ICU stay and roughly 1 liter BID was removed via the new PD catheter. Transfer from the ICU to the floor occurred on POD 1, where he was maintained on telemetry with no acute events. Discharge was on POD 13 after a course of diuresis and medical optimization.

Discussion

Table 1: Prevalence of CHDs for Individuals of All Ages, Among Children (0–17 Years of Age), and Among Adults (≥18 years of Age) by Severity and Race-Ethnicity in the United States, 2010.[8]

Category and Age Group	CHD Severity/Race-Ethnicity	Estimated US Prevalence per 1000 (95% Confidence Interval), %	Estimated No. of Individuals (95% Confidence Interval)*
CHD severity			
All ages	Overall	7.85 (7.79–7.92)	2 425 000 (2 405 000–2 444 000)
	Severe	0.92 (0.90–0.94)	283 000 (277 000–290 000)
Children	Overall	13.21 (13.03–13.39)	980 000 (966 000–993 000)
	Severe	1.66 (1.60–1.73)	123 000 (119 000–128 000)
Adults	Overall	6.16 (6.10–6.22)	1 444 500 (1 431 000–1 459 000)
	Severe	0.68 (0.66–0.70)	160 000 (155 000–165 000)
Race-ethnicity			
Children	Non-Hispanic white	13.31 (13.12–13.49)	620 000 (612 000–629 000)
	Non-Hispanic black	12.69 (12.50–12.88)	133 000 (131 000–135 000)
	Hispanic	13.26 (13.08–13.45)	227 000 (224 000–230 000)
Adults	Non-Hispanic white	6.36 (6.29–6.42)	1 104 000 (1 094 000–1 115 000)
	Non-Hispanic black	5.63 (5.56–5.69)	155 000 (153 000–156 000)
	Hispanic	5.58 (5.52–5.65)	186 000 (184 000–188 000)

CHD indicates congenital heart defect.
*Rounded to the nearest 1000; any minor inconsistencies between Tables 1 and 2 have resulted because estimates are the rounded mean values from 10 000 simulations.

Figure 1: Prevalence/Incidence of Congenital Heart Disease.[9]

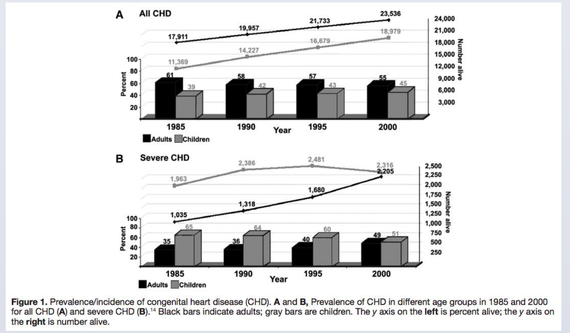


Figure 1. Prevalence/incidence of congenital heart disease (CHD). **A** and **B,** Prevalence of CHD in different age groups in 1985 and 2000 for all CHD (**A**) and severe CHD (**B**).[14] Black bars indicate adults; gray bars are children. The *y* axis on the **left** is percent alive; the *y* axis on the **right** is number alive.

In this case of a PD catheter placement with Mr. X, the anesthesia team extensively discussed with the surgical team regarding goals of care. Because this patient would not tolerate positive pressure ventilation without certain cardiovascular instability, the surgical team decided to perform this surgery open instead of laparoscopically.

Intraoperative Management Plan:

- Constant communication between anesthesia and surgical services
- Maintaining spontaneous ventilation to avoid positive pressure ventilation, which could lead to decreased CO
- Minimizing opioids to maintain adequate respiratory drive
- Maintaining baseline preoperative SpO2s and blood pressures
- Careful hemodynamic monitoring via arterial line, which was placed pre-induction
- Use of norepinephrine and vasopressin due to reduced EF of 40% and advanced CHD; avoidance of medications that increase PVR (e.g. Phenylephrine)
- Preventing air embolization with 22 micron filters given the patient's cyanotic defect

Conclusions

- Patients with congenital heart defects are increasingly likely to survive to adulthood, and this adult population is rapidly growing thanks to advancing surgical and medical management
- The demand for non-cardiac surgeries in CHD patients would also be expected to increase
- Anesthesiologists may not have much exposure to, or comfort in, managing adult CHD patients in the setting of non-cardiac surgery; this population is also at high risk for morbidity perioperatively
- Adequate anesthesia preoperative workup and perioperative management are critical in this high-risk population
- Establishing a plan of care with the surgical team may change the surgical plan, optimize postoperative outcomes, and prevent adverse events

References

1. Afilalo J, Therrien J, Pilote L, Ionescu-Ittu R, Martucci G, Marelli AJ. Geriatric congenital heart disease: burden of disease and predictors of mortality. J Am Coll Cardiol. 2011;58:1509-1515.
2. Marelli AJ, Ionescu-Ittu R, Mackie AS, Guo L, Dendukuri N, Kaouache M. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. Circulation. 2014;130:749-756.
3. Krasuski RA and Bashore TM. Congenital Heart Disease Epidemiology in the United States. Circulation. 2016;134:110–113. DOI: 10.1161/CIRCULATIONAHA.116.023370
4. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation. 2007;115:163- 172.
5. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2008;52:e143-e263.
6. Maxwell BG, Williams GD, Ramamoorthy C. Knowledge and attitudes of anesthesia providers about noncardiac surgery in adults with congenital heart disease. Congenit Heart Dis. 2013;9:45-53.
7. Maxwell BG, Posner KL, Wong JK, et al. Factors contributing to adverse perioperative events in adults with congenital heart disease: a structured analysis of cases from the Closed Claims Project [published online May 28, 2014]. Congenit Heart Dis. doi:10.1111/chd.12188.
8. Gilboa SM, Devine OJ, Kucik JE, et al. Congenital Heart Defects in the United States; Estimating the Magnitude of the Affected Population in 2010. Circulation. 2016;134:101–109. DOI: 10.1161/CIRCULATIONAHA.115.019307
9. Stout KK et al. Chronic Heart Failure in Congenital Heart Disease: A Scientific Statement from the American Heart Association. Circulation, 2016, DOI: 10.1161/CIR.0000000000000352

Acknowledgements

- The authors would like to thank the Emory University Department of Anesthesiology for their support.