Pacing the Way: Challenges of Pacemakers in Pediatrics and Adults with Congenital Heart Disease

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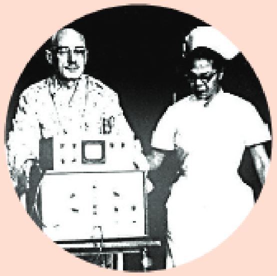
Cleveland Clinic Children's

Disclosure

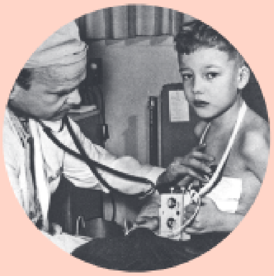
- I have no financial relationships to disclose relevant to this presentation

Paradigm Shifts in Cardiac Pacemakers

1950s
AC-powered pacemakers tethered to an extension cord (Furman)



1950s
Battery-powered transistorized "wearable" pacemakers (Lillehei/Bakken)

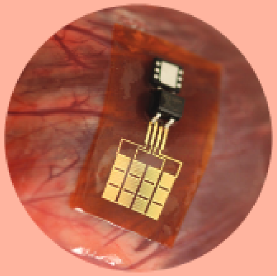


1958
First fully implantable pacemaker (Elmqvist/Senning)

2015
Implantable pacemaker—basic system had not evolved significantly

2016
Leadless pacemaker—the entire device is placed within cardiac chambers

Future
Batteryless devices, which harvest cardiac motion to power pacing circuits



Mulpuru, S.K. et al. J Am Coll Cardiol. 2017;69(2):189-210.

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Objectives

- Indications for pacing in pediatrics and adults with congenital heart disease (ACHD)

- Complexities of pacing in pediatrics and ACHD

  - How to choose type of pacemaker
    - Transvenous vs. epicardial vs. leadless

  - Specific ACHD cases

Indications

- Sinus node dysfunction
  - Common in Fontan patients and patients status post atrial switch procedure
- Advanced second or third degree AV block
  - Congenital
  - Postoperative
- Prevention or treatment of tachyarrhythmia
- Cardiac resynchronization therapy in case of dysfunction

Factors to Consider

- Size of the patient (age and weight)
- Cardiac and venous anatomy
- Surgical procedures performed that can affect the access to certain cardiac structures
- Number of previous surgical procedures
- Need for future magnetic resonance imaging

Epicardial Pacemaker

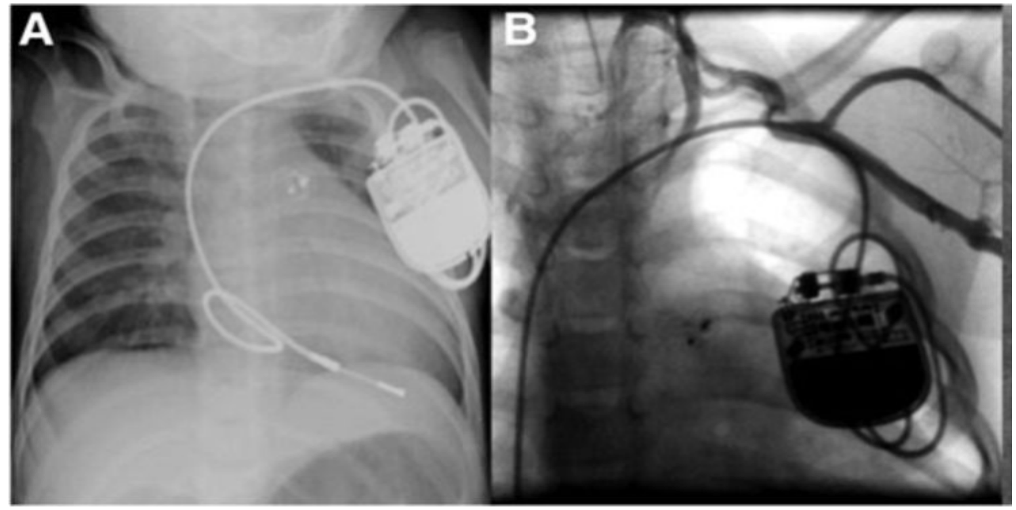
When to consider

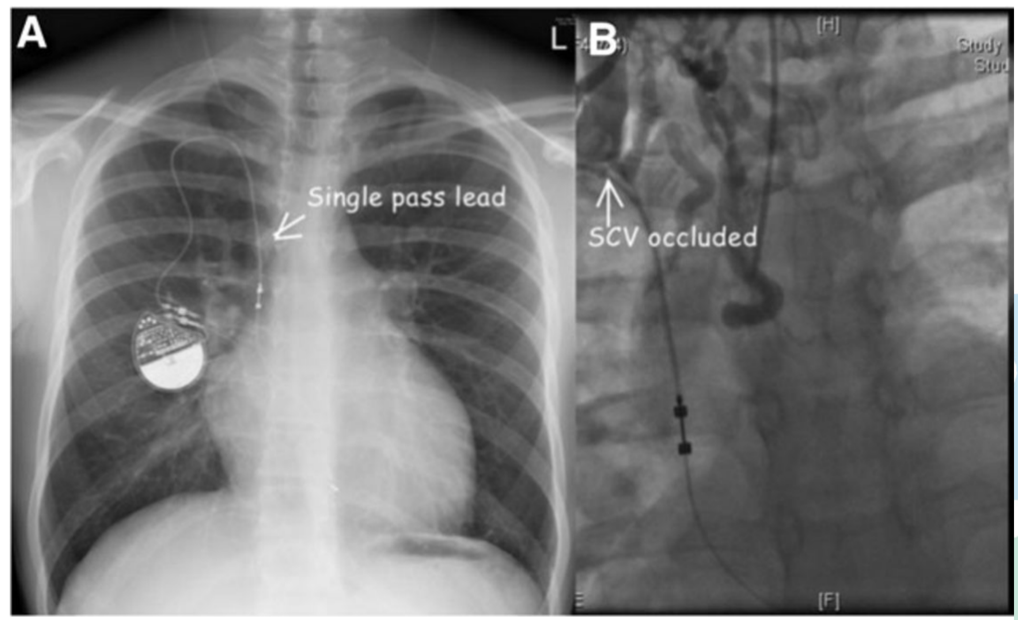
- Patients < 15 kg
- Presence of intracardiac shunt lesion
- Limited access to atrium and ventricle
  - Single ventricle patients post Fontan
- Need for cardiac surgery

Disadvantages

- Need for cardiac surgery
- Higher chronic stimulation threshold
- Higher lead failures and fractures, and
- Early depletion of battery life
- Coronary artery compression

Risk of venous occlusion is as high as **77%** in < 5 kg and 13% in > 5 kg





Konta et al. Circ EP 2016

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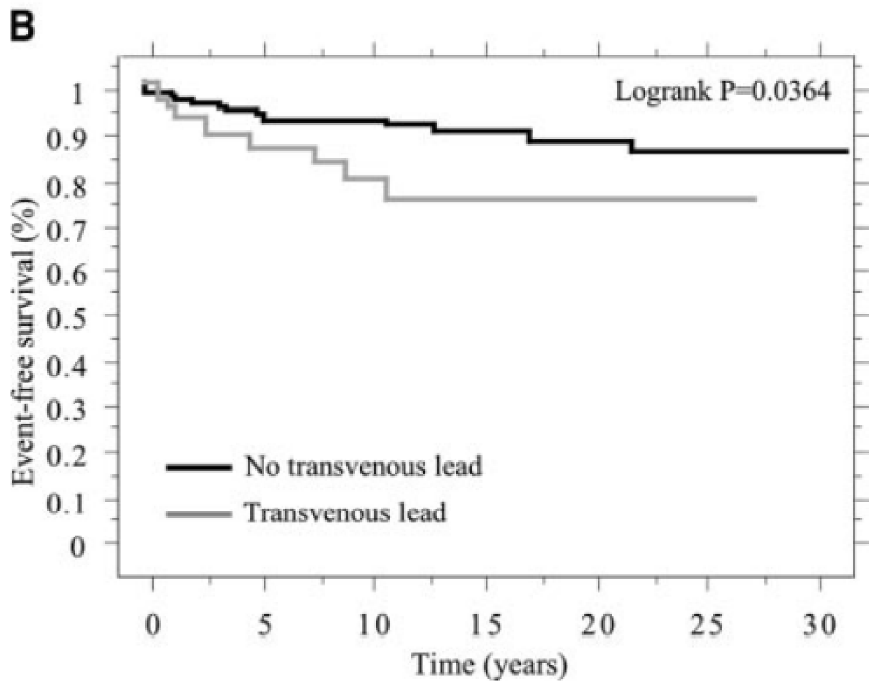
- Multicenter, retrospective cohort of 202 patients with intracardiac shunts:
  1. 64 with transvenous (TV) leads
  2. 56 with epicardial leads
  3. 82 with right to left shunts but no pacemaker or ICD leads

- Outcome: occurrence of systemic thromboemboli

- **Presence of TV leads was an independent predictor of outcome with greater than 2-fold increased risk.**



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Khairy P et al. *Circulation*. 2006

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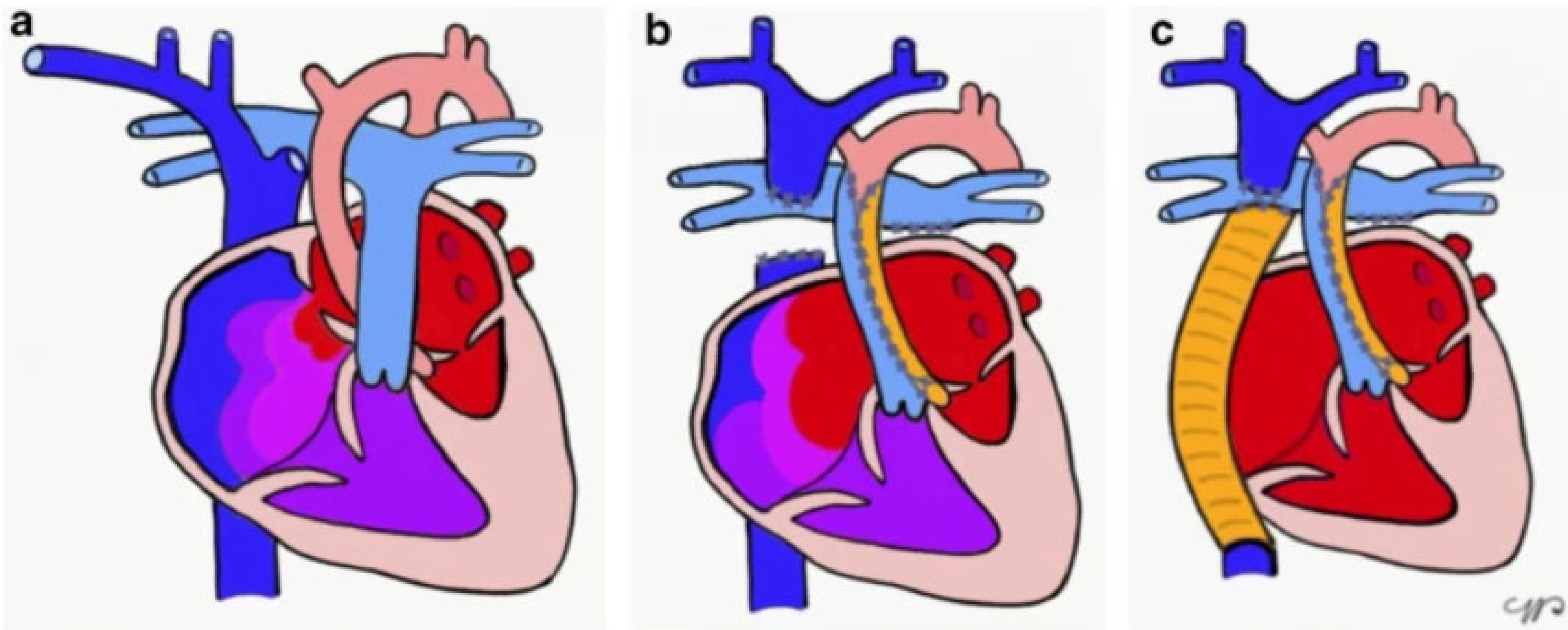

Perucca et al. Pediatric Radiology 2021

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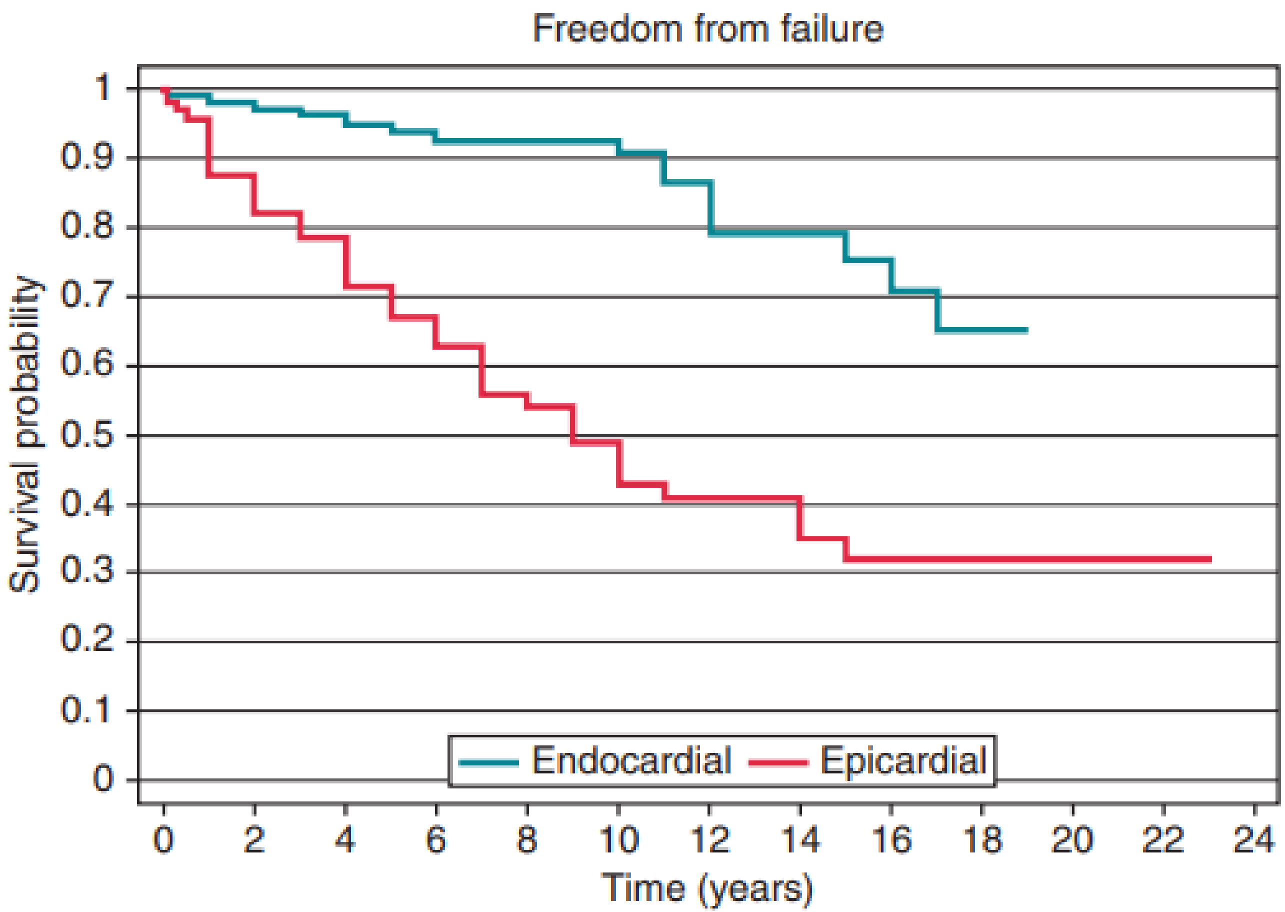

Number at risk

Endocardial	117	105	89	76	61	50	35	25	17	5	0	0	0
Epicardial	170	126	100	65	37	25	16	14	10	5	5	2	0

M.S. Silvetti et al. Europace. 2013

Epicardial Pacemaker

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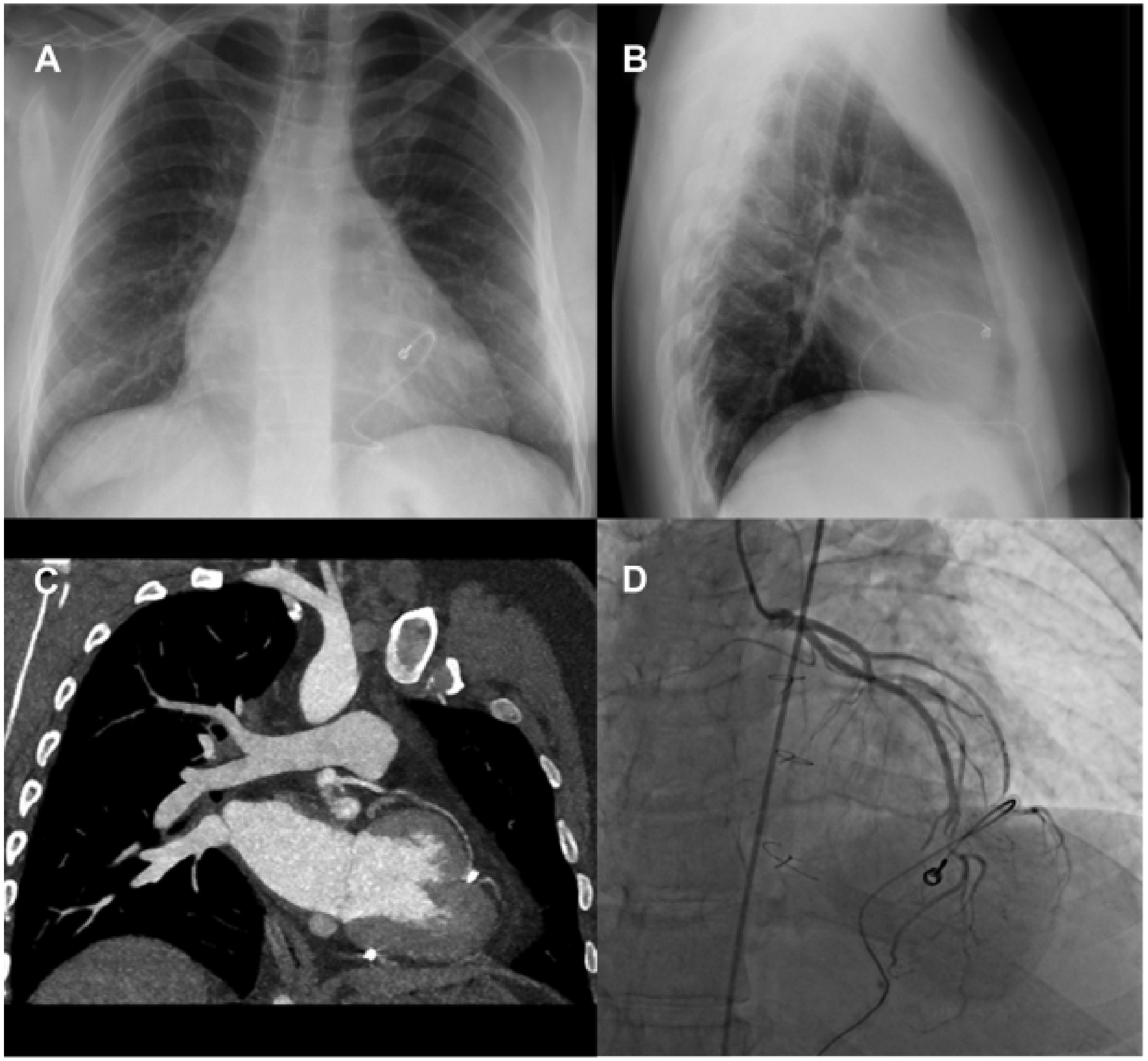


Mah D et al. Heart Rhythm 2021

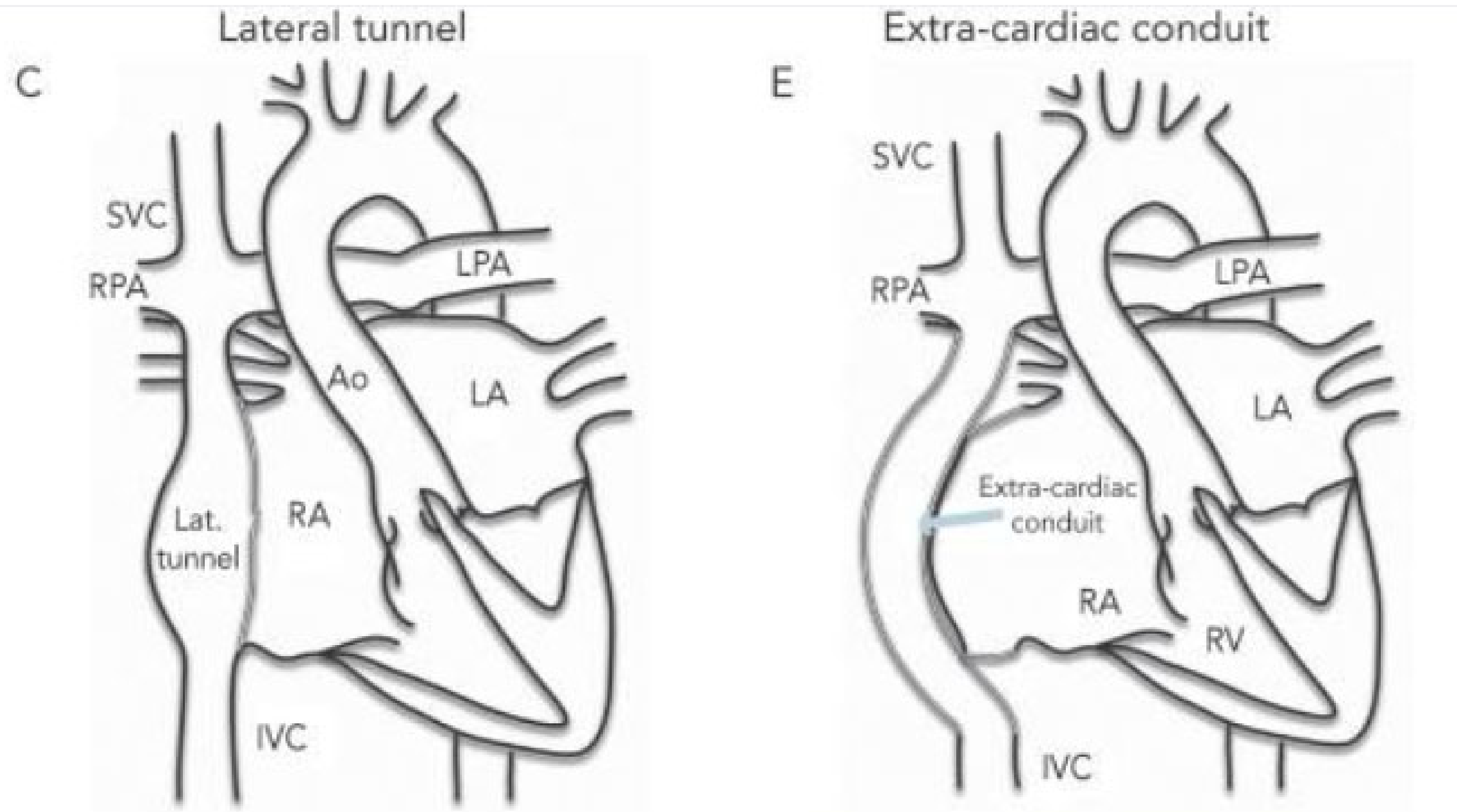
Endocardial Pacemaker

What & when to consider

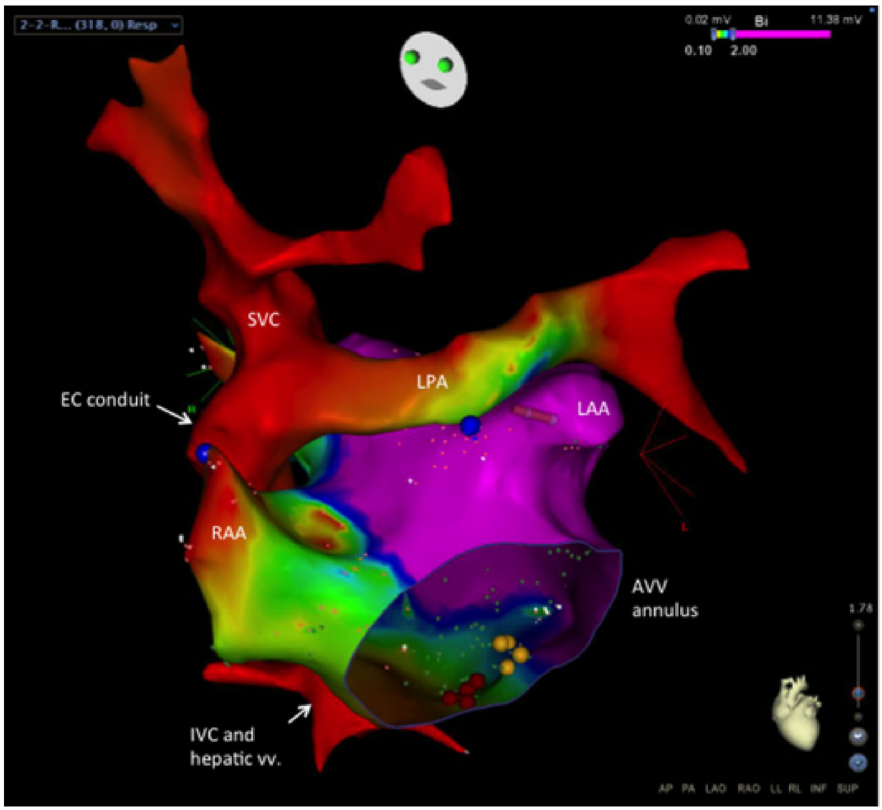
- Patient > 15 kg
- Presence of intracardiac shunt lesion
- Know the patient's anatomy and surgical history!!!

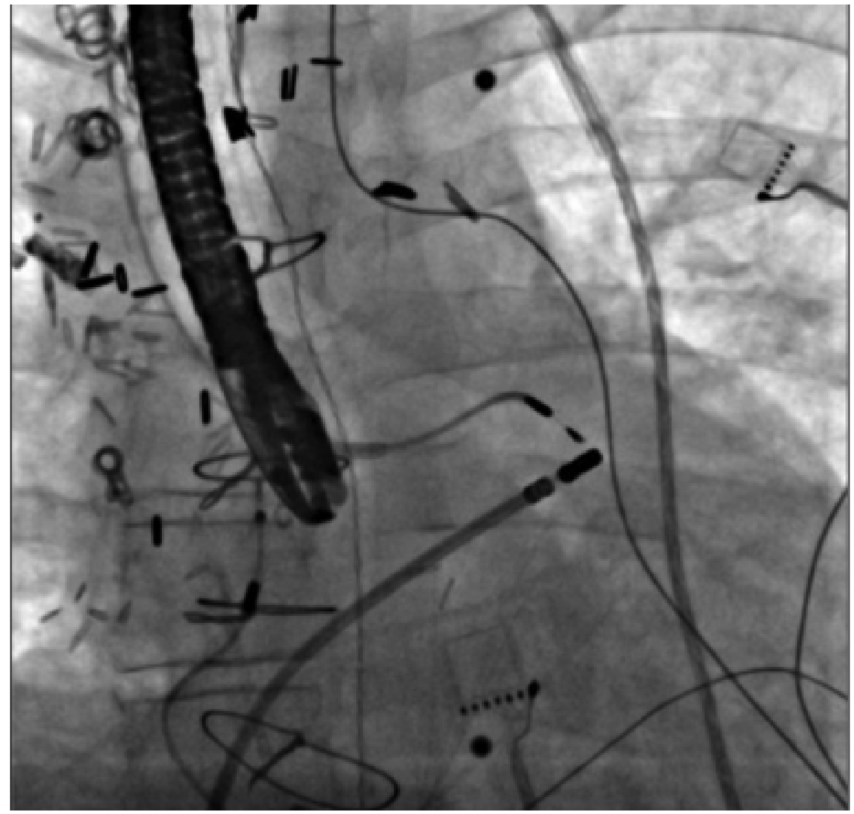
Disadvantages

- Vascular occlusion
- Thromboembolism risk
- Need for extraction



Chubb H et al. AER 2016

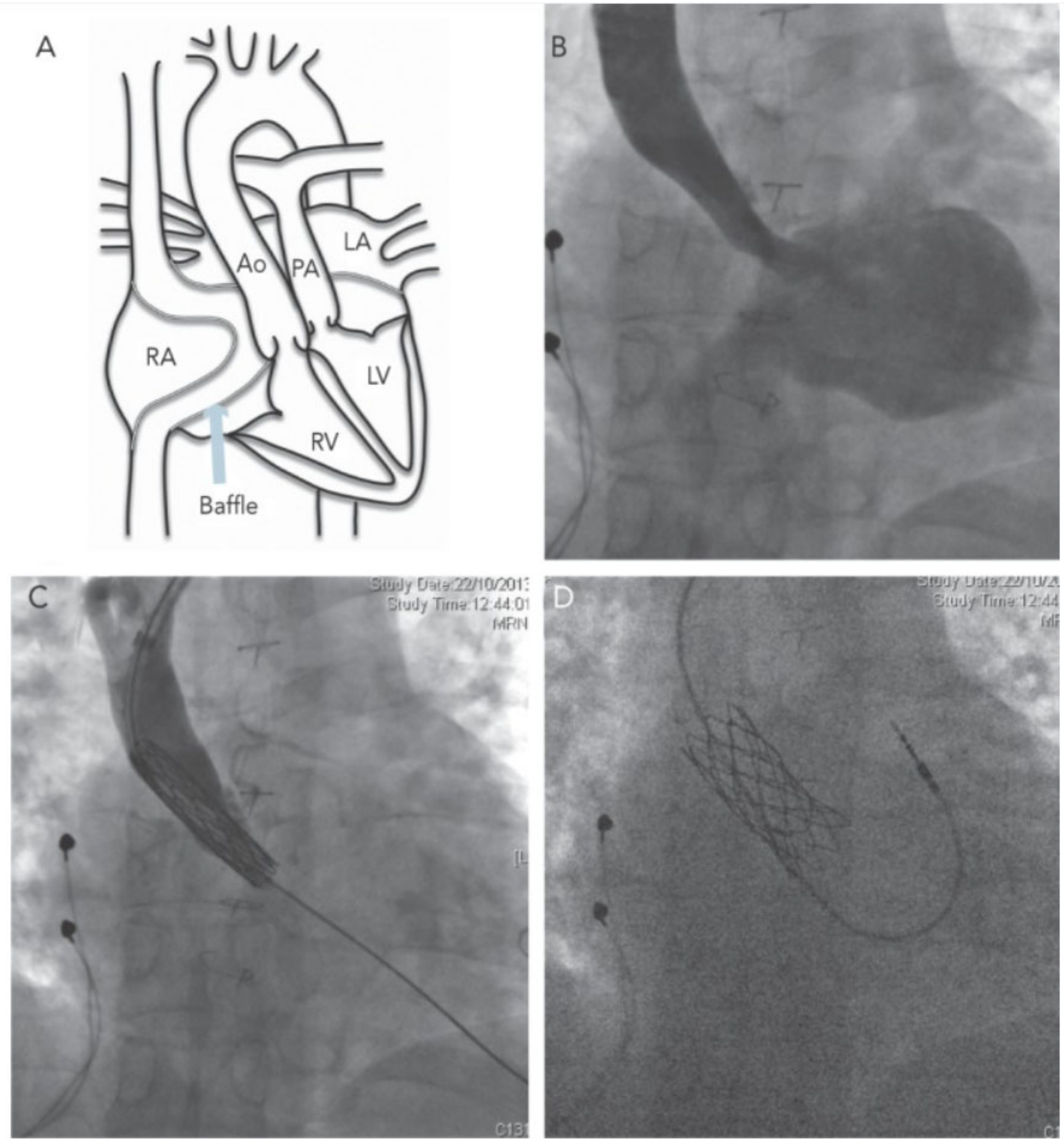




Moore J et al. JCE 2014

Atrial Switch Procedures

- High incidence of baffle obstruction
- Risk of phrenic nerve stimulation w/ LA lead
- Smooth LV wall
- CS anatomy



Endocardial Pacemaker

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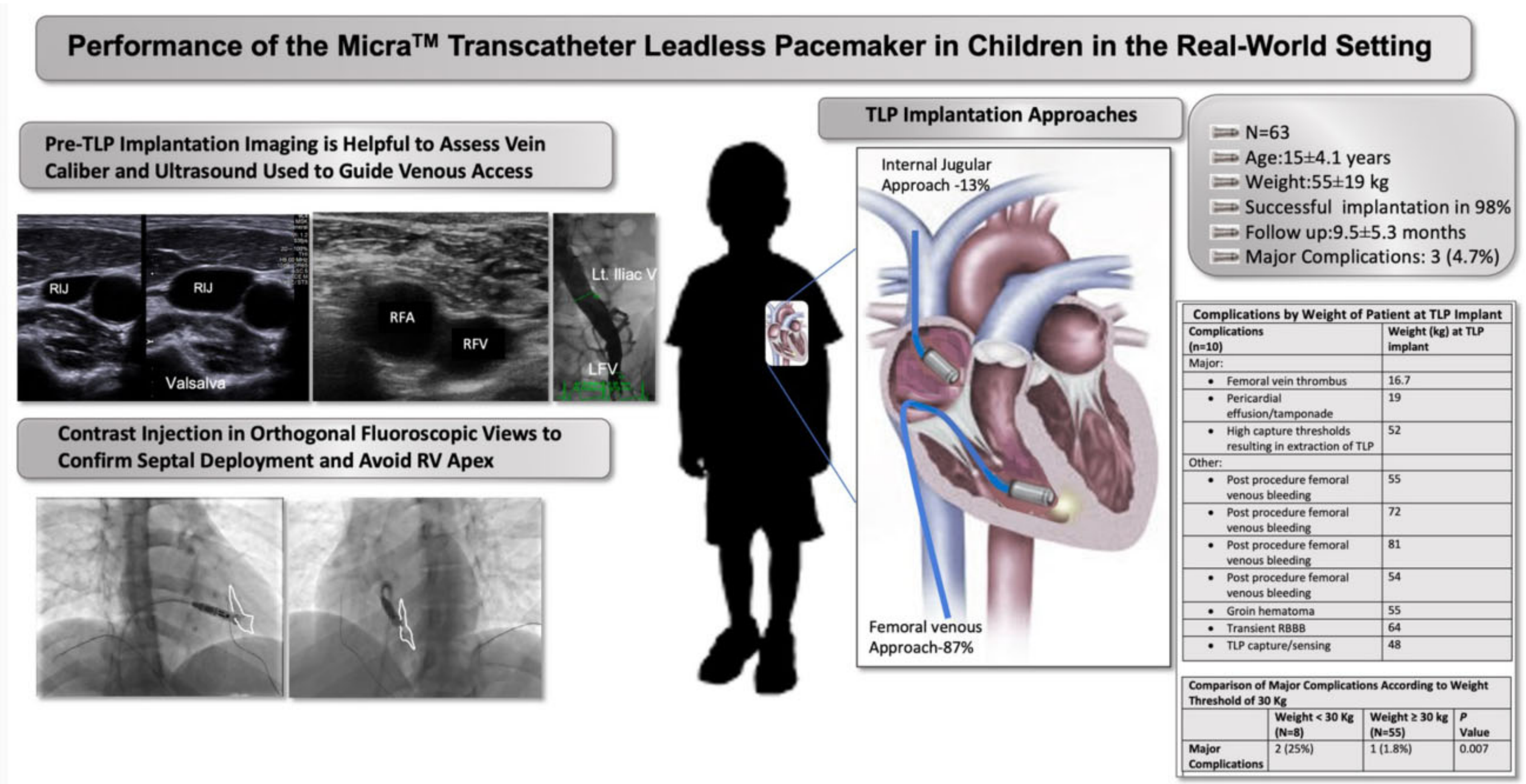
Leadless Pacemaker



Complications by Weight of Patient at TLP Implant

Complications (n=10)	Weight (kg) at TLP implant
Major:	
• Femoral vein thrombus	16.7
• Pericardial effusion/tamponade	19
• High capture thresholds resulting in extraction of TLP	52
Other:	
• Post procedure femoral venous bleeding	55
• Post procedure femoral venous bleeding	72
• Post procedure femoral venous bleeding	81
• Post procedure femoral venous bleeding	54
• Groin hematoma	55
• Transient RBBB	64
• TLP capture/sensing	48

Comparison of Major Complications According to Weight Threshold of 30 Kg

	Weight < 30 Kg (N=8)	Weight ≥ 30 kg (N=55)	*P* Value
Major Complications	2 (25%)	1 (1.8%)	0.007

Shah M et al. Circ EP 2022

Current Challenges with Leadless Pacing

- Access
- AV synchrony
- Adequate sensing and capture in patients with large VSD patch
- Extractability?
- Long term impact of multiple leadless pacemakers on ventricular myocardium

30 yo w/ TOF status post repair with severe BIV dysfunction (RV EF 27%, LV EF 24%), CRBBB, and NSVT. On maximal GDT.

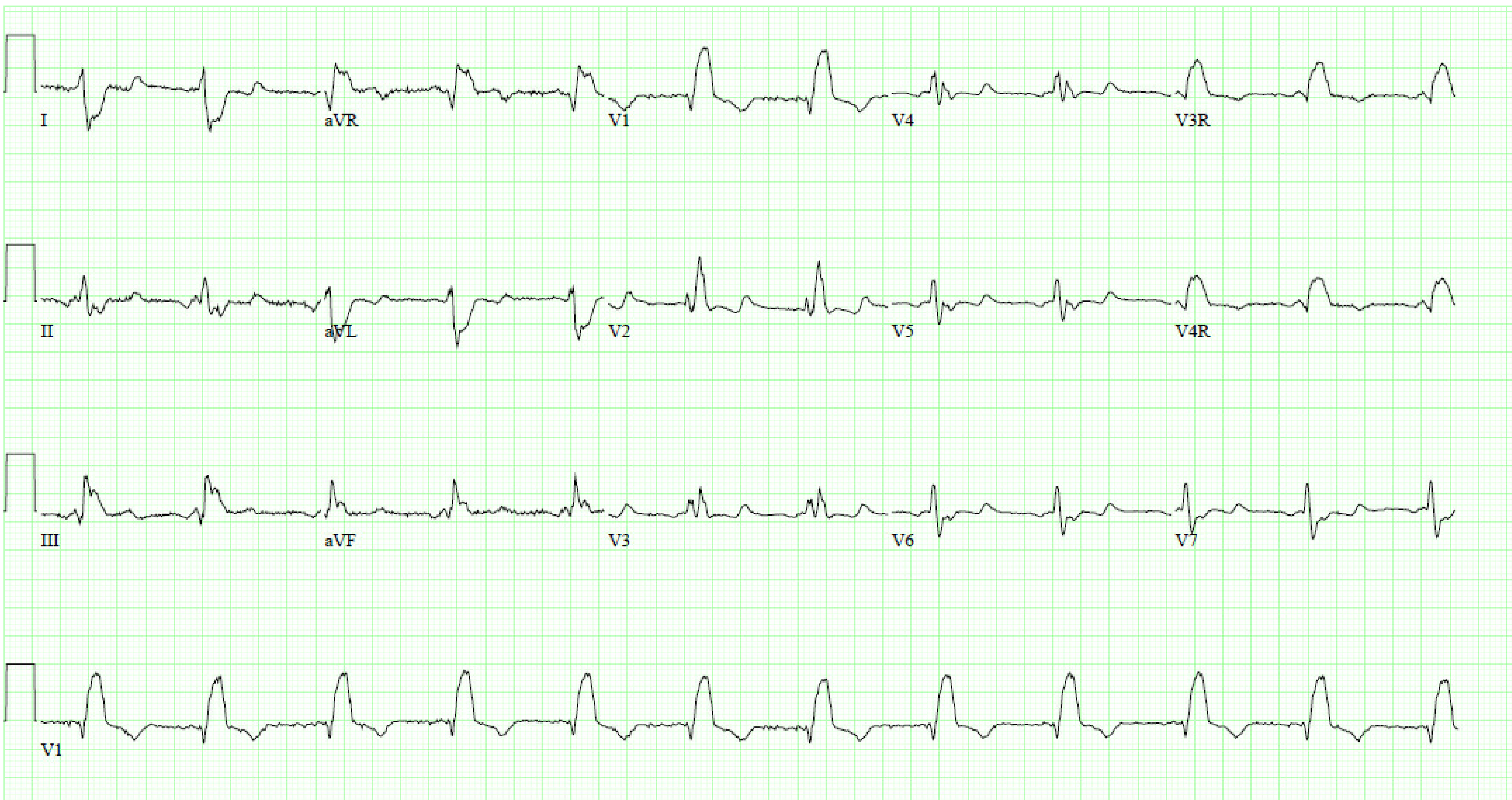
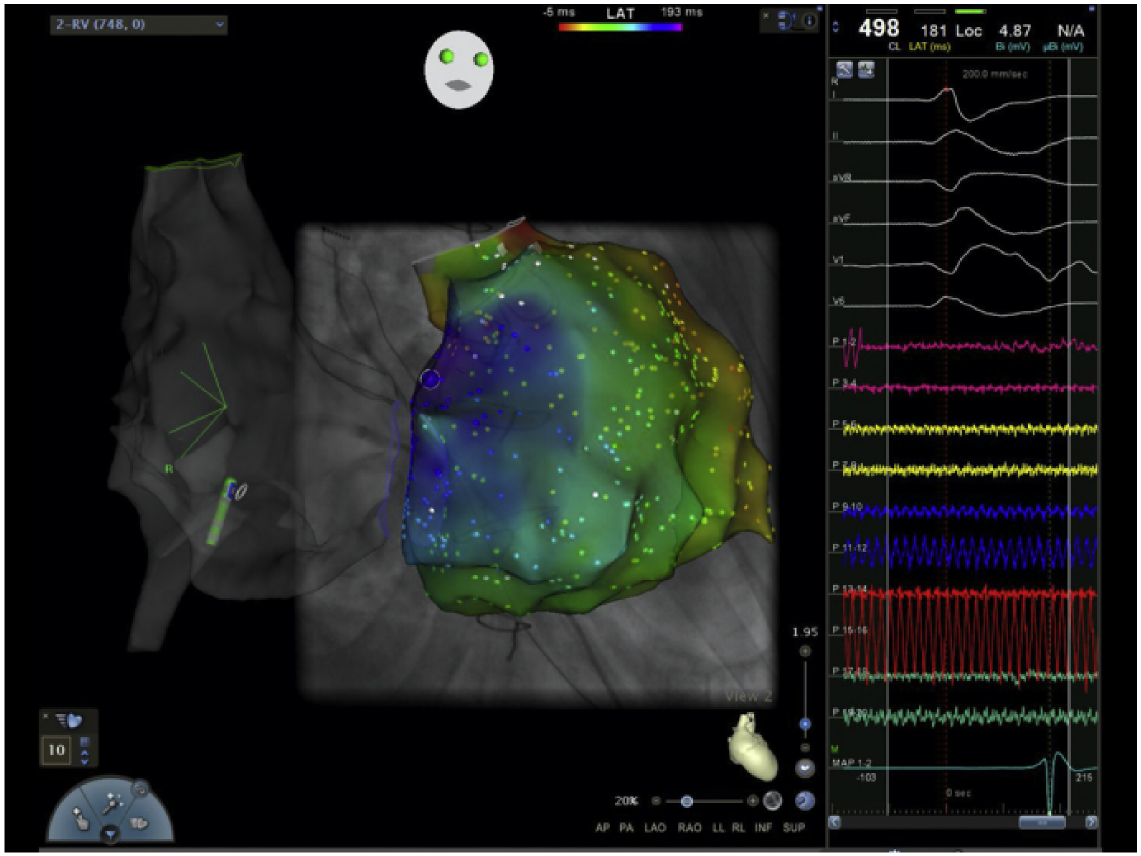
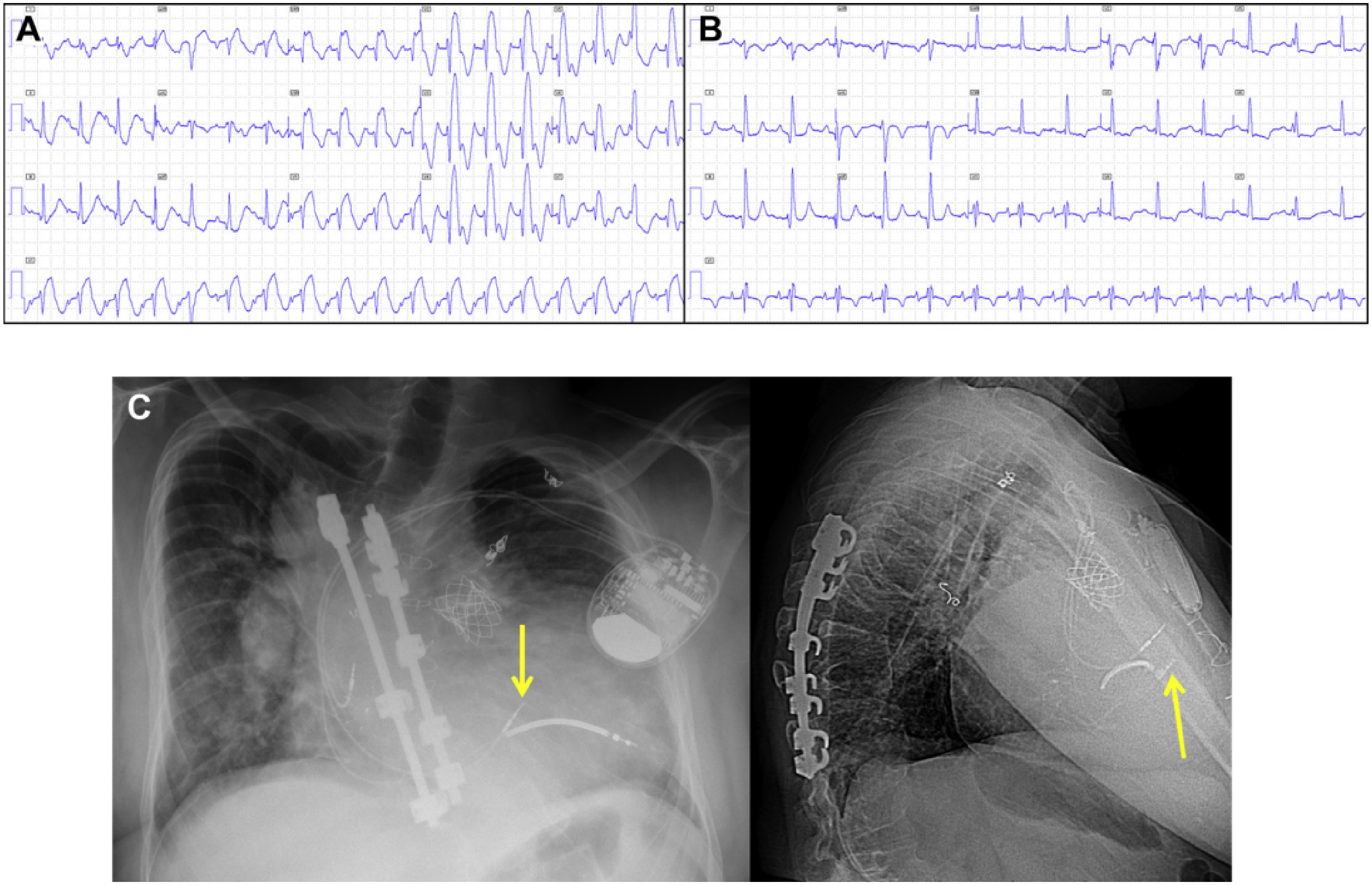


FIGURE 1 Representative Electroanatomic Map of Right Ventricular Activation in Our Patients With Tetralogy of Fallot



Mah D et al. JACC EP 2021

FIGURE 5 Representative Electrocardiograms and Chest X-Rays for CRT of a Subpulmonary Right Ventricle in a Patient With Tetralogy of Fallot



Mah D et al. JACC EP 2021

Conclusions

- Device therapy in children and adults with CHD poses unique challenges
- One-size-fits-all approach does **NOT** apply to this population
- Important to be aware of their anatomy and surgical procedures they have had

Questions?

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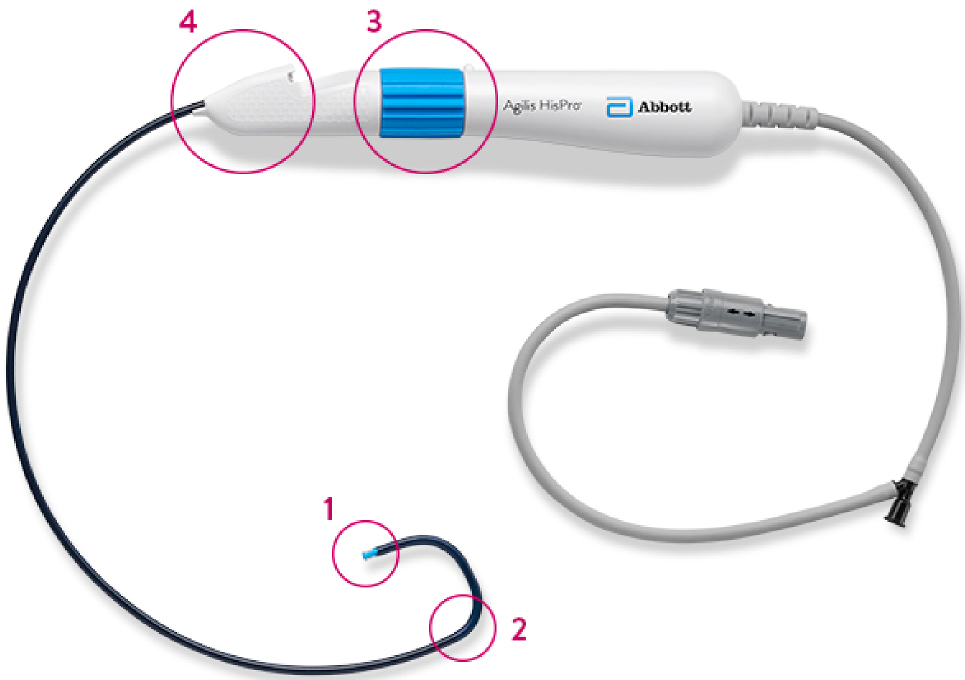
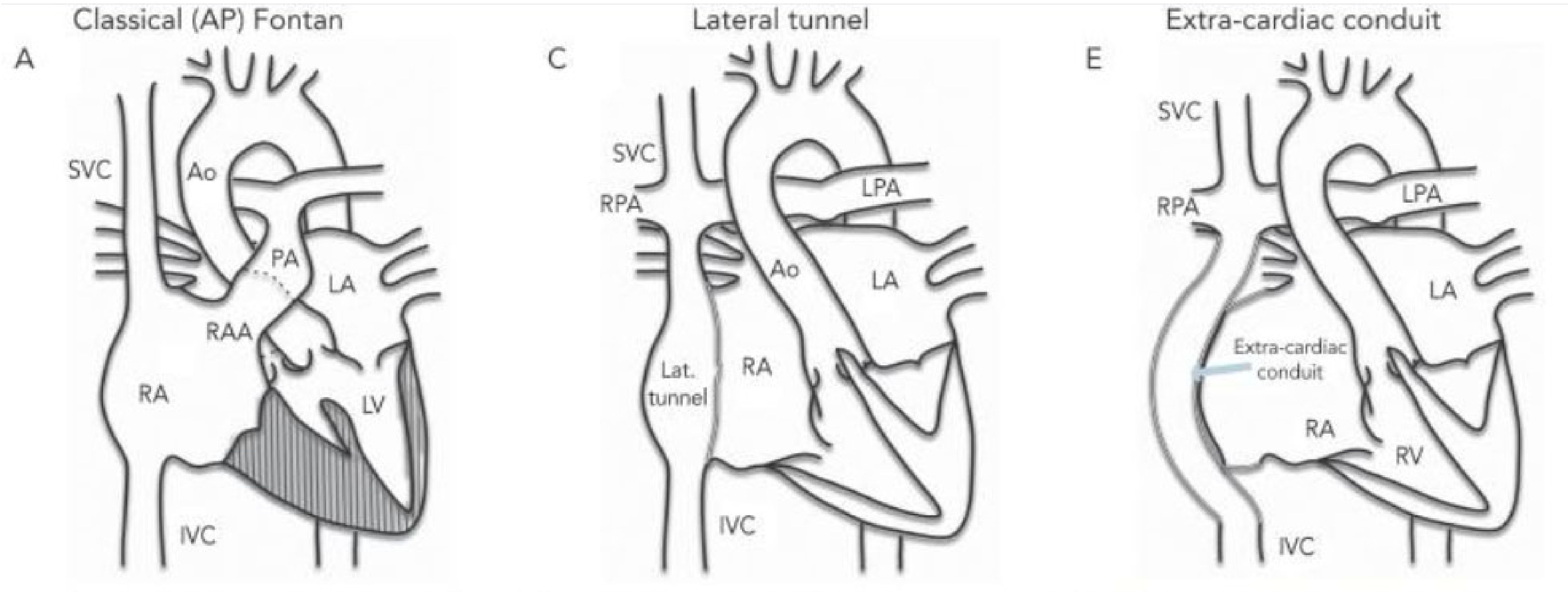
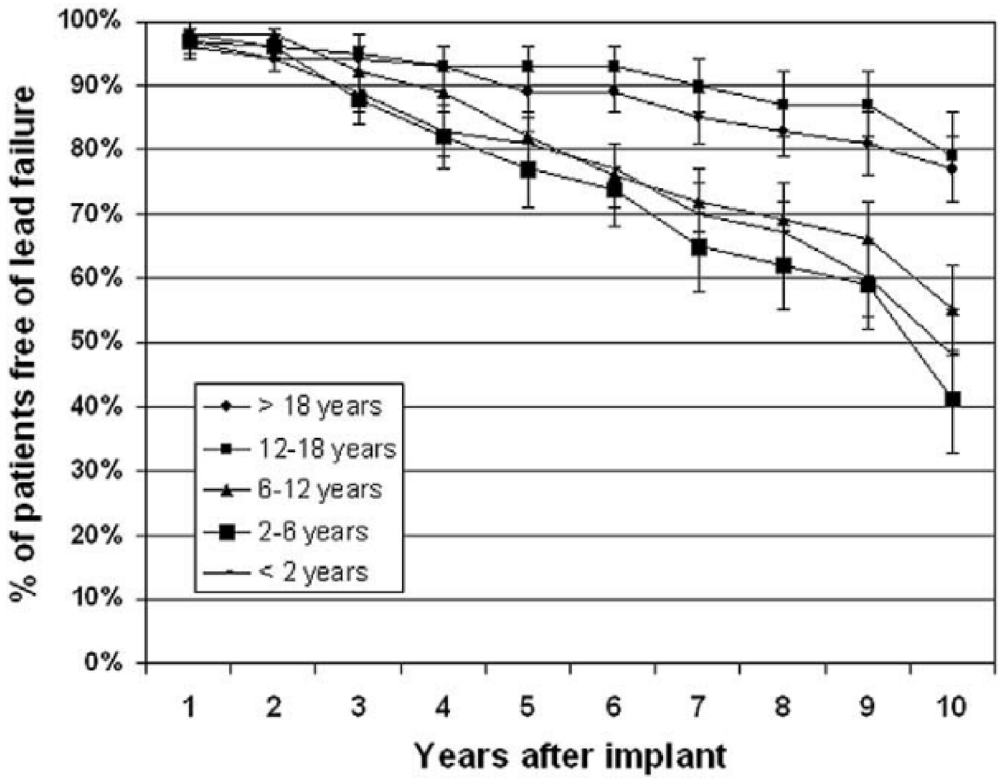
Every life deserves world class care.

- OD: 10.5 F, ID: 7.0 F
- Use with a minimum 10.5 F introducer
- Catheter designed for delivery of a 6F lead
- Working length of the catheter is 38 cm
- Requires at least a 58 cm lead length



1. Integrated bipolar tip electrodes
2. Dual deflection and curve
3. 180° deflection control knob


Classical (AP) Fontan
A
SVC
Ao
PA
LA
RAA
RA
LV
IVC
Lateral tunnel
C
SVC
RPA
LPA
Ao
LA
Lat.
tunnel
RA
IVC
Extra-cardiac conduit
E
SVC
RPA
LPA
LA
Extra-cardiac
conduit
RA
RV
IVC



100%
90%
80%
70%
60%
50%
40%
30%
20%
10%
0%
% of patients free of lead failure
1
2
3
4
5
6
7
8
9
10
Years after implant
> 18 years
12-18 years
6-12 years
2-6 years
< 2 years

TABLE 1 Patient Responses to Resynchronization Therapy

	Age at CRT	Sex	Anatomy	Variable	Pre-CRT	Post-CRT
Single RVs						
Patient #1	5 yrs	M	Right-dominant atrioventricular canal	QRS duration, ms	180	110
				RV function	Severe dysfunction	Moderate dysfunction
				BNP, pg/dl	3,312	114
Patient #2	3 yrs	F	Double-outlet RV, mitral stenosis, pulmonary atresia	QRS duration, ms	130	105
				RV function	Severe dysfunction	Moderate dysfunction
				BNP, pg/dl	613	52
Subpulmonary RVs						
Patient #3	27 yrs	M	Tetralogy of Fallot, pulmonary atresia	QRS duration, ms	250	140
				RV end-diastolic volume, ml	494	429
				RV ejection fraction, %	8	25
				LV end-diastolic volume, ml	159	152
				LV ejection fraction, %	33	61
				BNP, pg/dl	243	281
				Vo_2, ml/kg/min	12.6 (37% predicted)	12.1 (31% predicted)
Patient #4	36 yrs	M	Tetralogy of Fallot, pulmonary atresia	QRS duration, ms	260	160
				RV end-diastolic volume, ml	652	443
				RV ejection function, %	7	16
				LV ejection fraction, %	30	46
				BNP, pg/dl	40	43
				Weight, kg	113	99
Patient #5	48 yrs	M	Tetralogy of Fallot	QRS duration, ms	250	165
				RV function	Moderate dysfunction	Moderate dysfunction
				LV ejection fraction, %	51	57
LVs with right bundle branch block						
Patient #6	19 months	F	Multiple small left-sided structures (mitral and aortic stenosis, coarctation) with VSD	QRS duration, ms	130	95
				LV ejection fraction, %	26	55
Patient #7	7 months	F	Multiple small left-sided structures (mitral and aortic stenosis, coarctation) with VSD and hypoplastic left pulmonary artery	QRS duration, ms	115	90
				LV function	40%	Mild dysfunction