Rhythmologie

To Pace or not to Pace?

Jan Till

Royal Brompton

London

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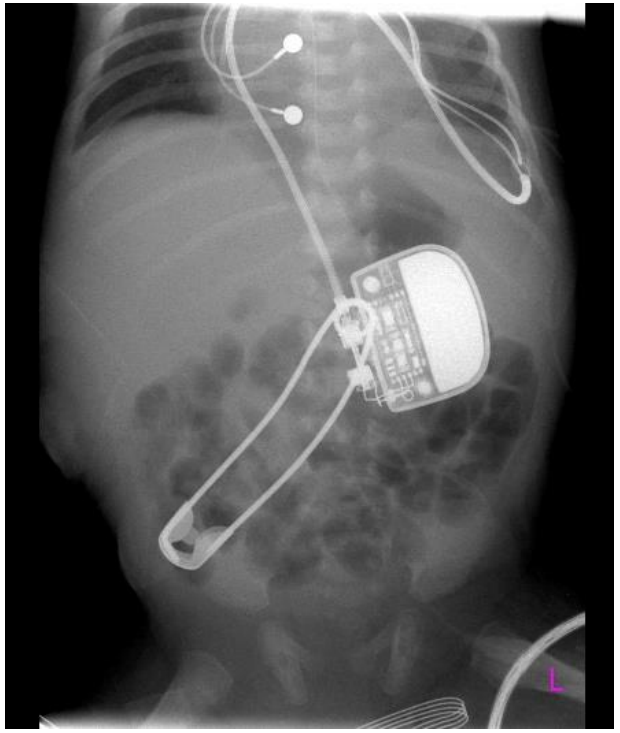
Why might we think about pacing a child or young person?

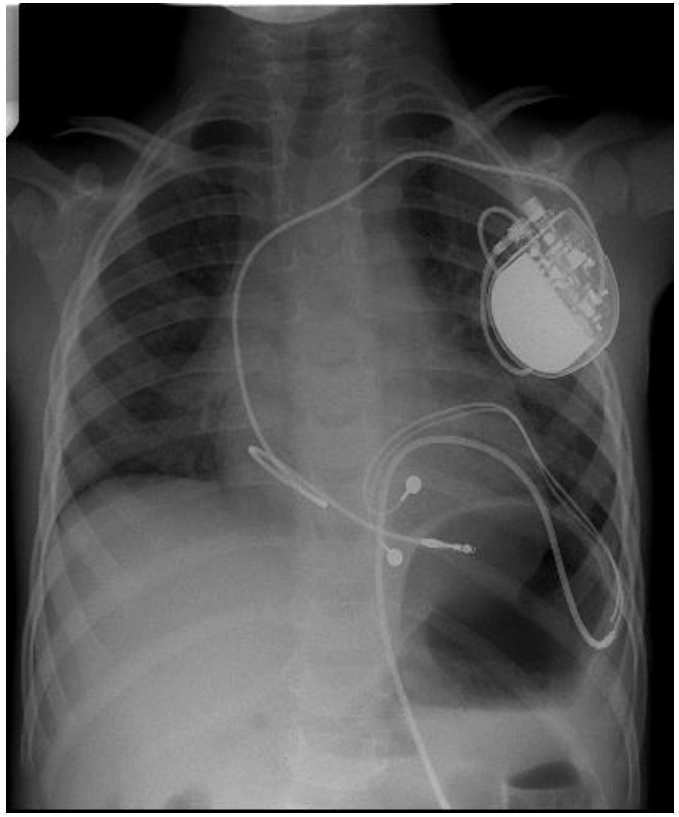
- AV block
- Sinus node disease
- Advanced heart failure
  - Congenital or acquired
  - Structurally normal heart
  - CHD +/- surgery

- Neuromuscular disease
- Myotonic muscular dystrophy
- Kearns Sayre syndrome
- Erb dystrophy (limb girdle) peroneal muscular atrophy
- Progressive conduction disorders
- RAS/neurocardiogenic syncope
- Long QT
- Congenital hypoventilation syndrome

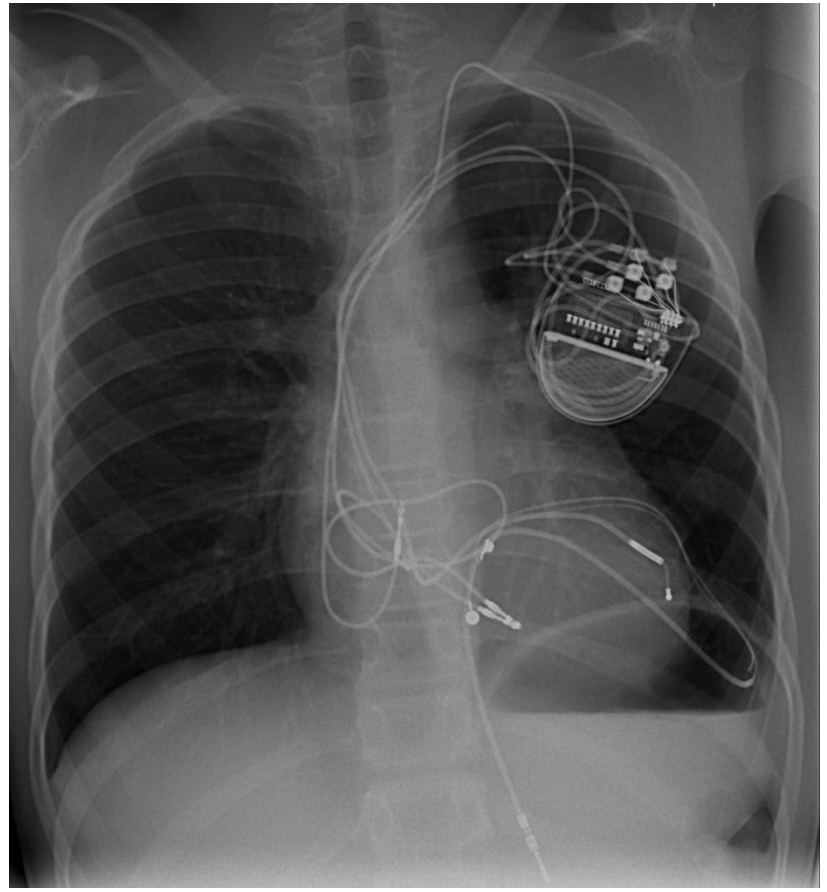
Congenital Complete AV Block



Preterm neonate 2.1 Kg



Age 2 yrs



Age 8 yrs

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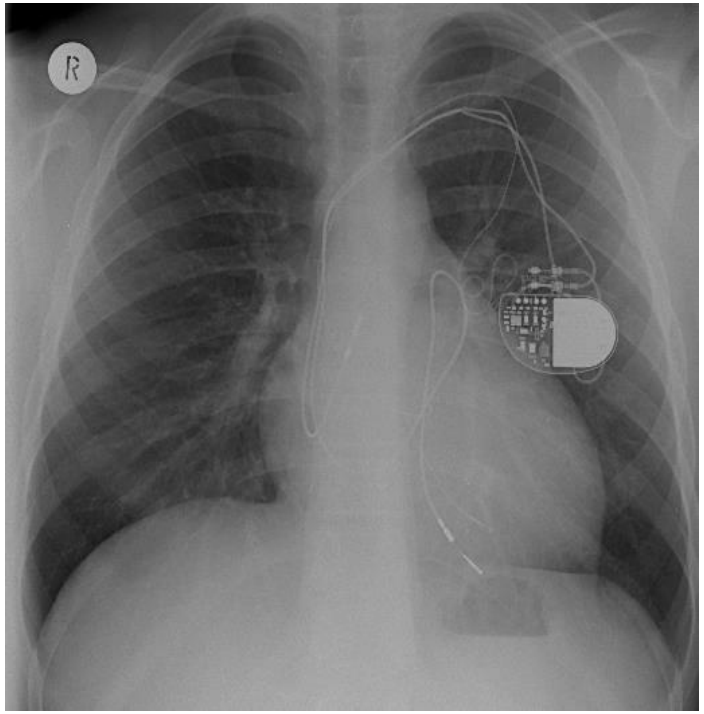

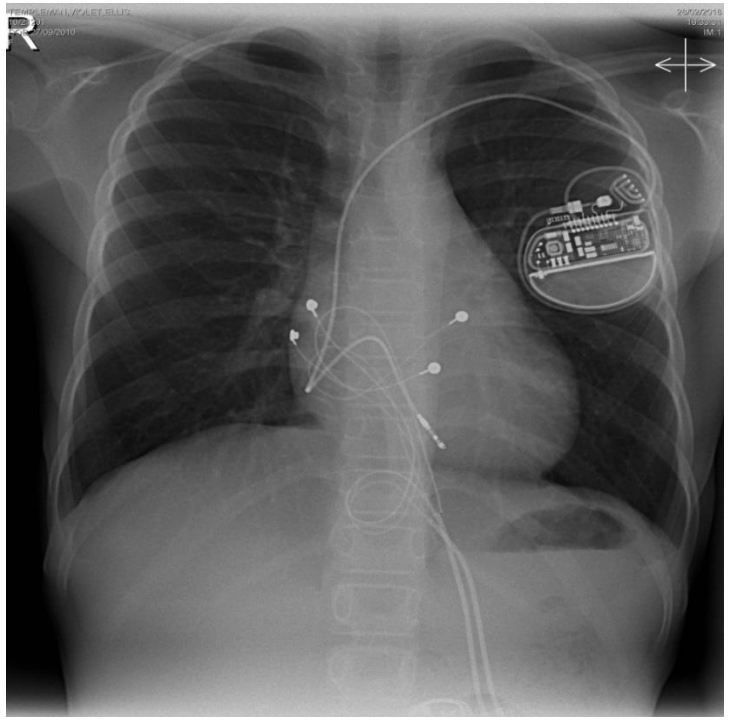

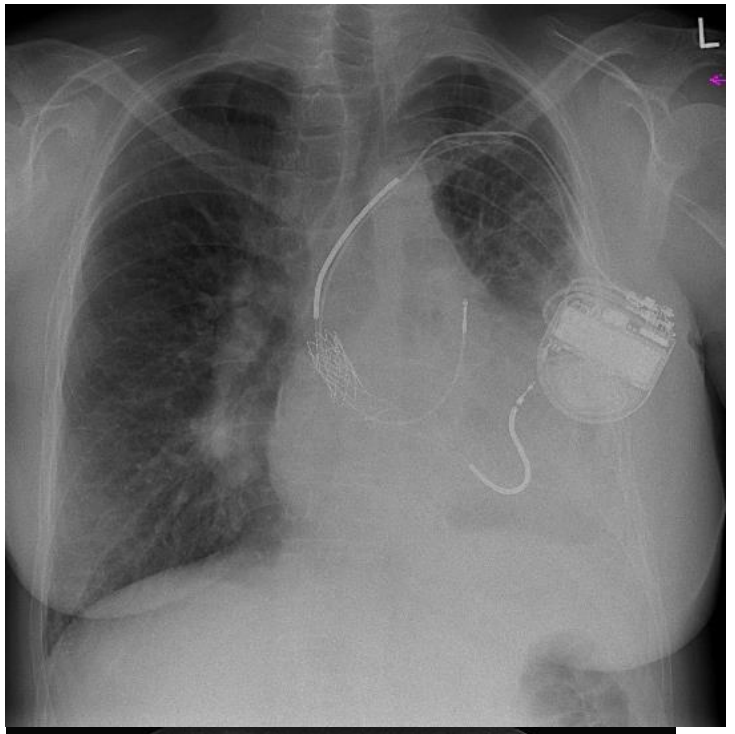

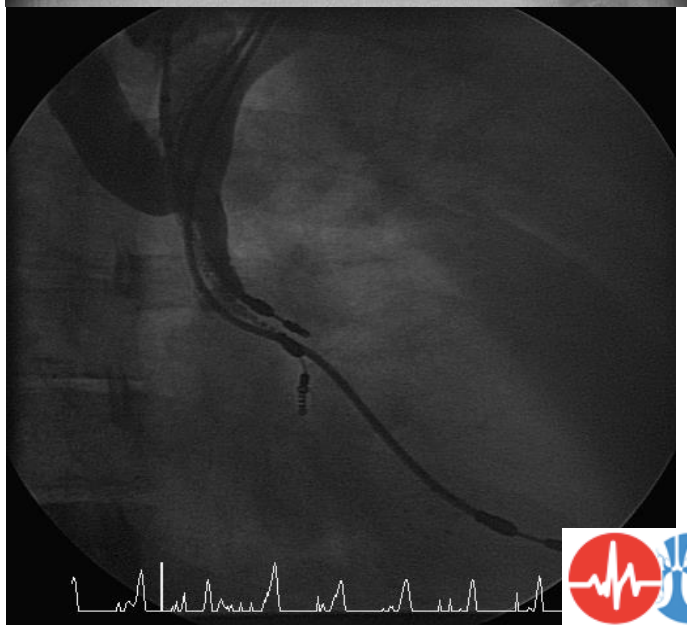

Challenges of pacing children

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Long term consequences of pacing early

Risk of condition

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Europace Advance Access published July 12, 2013

Europace
doi:10.1093/europace/eut082

EHRA/AEPC CONSENSUS STATEMENT

Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group joint consensus statement

Josep Brugada[1]*, Nico Blom[2], Georgia Sarquella-Brugada[3], Carina Blomstrom-Lundqvist[4], John Deanfield[5], Jan Janousek[6], Dominic Abrams[7], Urs Bauersfeld[8†], Ramon Brugada[9], Fabrizio Drago[10], Natasja de Groot[11], Juha-Matti Happonen[12], Joachim Hebe[13], Siew Yen Ho[14], Eloi Marijon[15], Thomas Paul[16], Jean-Pierre Pfammatter[17], and Eric Rosenthal[18]

	ESC guidelines 2013	ACCF/AHA/HRS guidelines 2008 updated 2012
Congenital AV block		
Symptomatic advanced second- or third-degree AV block	Class I, level C	Class I, level C
Asymptomatic high degree AV block with ventricular dysfunction	Class I, level C	Class I, level B
Asymptomatic high degree AV block with prolonged QTc interval	Class I, level C	–
Asymptomatic high degree AV block with complex ventricular ectopy	Class I, level C	Class I, level B
Asymptomatic high degree AV block with wide QRS escape rhythm	Class I, level C	Class I, level B
Asymptomatic high degree AV block with abrupt ventricular pauses >threefold the basic cycle length	Class I, level C	Class IIa, level B
Asymptomatic third-degree AV block in the infant with a ventricular rate <55 bpm or with CHD and a ventricular rate <70 bpm	–	Class I, level C
Third-degree AV block beyond the first year of life with an average heart rate <50 bpm	–	Class IIa, level B
Asymptomatic high degree AV block with a ventricular rate <50 bpm	Class I, level C	–
Third-degree AV block beyond the first year of life with symptoms due to chronotropic incompetence	–	Class IIa, level B
High degree AV block in asymptomatic children/adolescents in absence of the above risk conditions	Class IIb, level C	Class IIb, level B
Asymptomatic type I second-degree AV block	–	Class III, level C
Postoperative AV block		
Postoperative advanced second- or third-degree AV block that persists >7 days after cardiac surgery (10 days in ESC guidelines)	Class I, level B	Class I, level B
Transient postoperative third-degree AV block that reverts to sinus rhythm with residual bifascicular block	Class IIa, level C	Class IIb, level C
Unexplained syncope in the patient with prior CHD surgery complicated by transient complete heart block with residual fascicular block	–	Class IIa, level B
Transient postoperative AV block with return of normal AV conduction in the otherwise asymptomatic patient	–	Class III, level B
Asymptomatic postoperative bifascicular block with/without first-degree AV block in the absence of prior transient complete AV block	–	Class III, level C

Baruteau Eur J Pediatrics 2016

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Level of evidence

A data derived available from multiple randomised trials or meta-analyses

B data derived from single randomised clinical trial or large non randomised studies

C consensus of opinion of experts and/or data derived from small studies retrospective studies or registries

Eur Heart J. 1981 Aug;2(4):281-8.

Congenital complete heart block in adolescence and adult life. A follow-up study.

Esscher EB.

Pediatrics. 1982 Jun;69(6):728-33.

Diagnosis, management, and long-term results of patients with congenital complete atrioventricular block.

Pinsky WW, Gillette PC, Garson A Jr, McNamara DG.

N Engl J Med. 1987 Apr 2;316(14):835-9.

Use of ambulatory electrocardiographic monitoring to identify high-risk patients with congenital complete heart block.

Dewey RC, Capeless MA, Levy AM.

Am Heart J. 1989 Dec;118(6):1193-8.

Congenital complete heart block in patients without anatomic cardiac defects.

Sholler GF[1], Walsh EP.

Pacing Clin Electrophysiol. 1997 Aug;20(8 Pt 2):2098-101.

Natural history of congenital complete atrioventricular block.

Michaelsson M[1], Riesenfeld T, Jonzon A.

Disorders of AV Conduction
Complete congenital atrioventricular block

- Class I
  - Complete congenital atrioventricular block in a newborn or an infant with a ventricular rate < 55 bpm or with CHD and a ventricular rate <70 bpm (C)
  - Complete congenital atrioventricular block with a wide complex escape rhythm, complex ventricular ectopy, or ventricular dysfunction (B)
  - Complete congenital atrioventricular block beyond first year of life with an average heart rate <50 bpm, abrupt pauses in ventricular rate 2-3x basic cycle length or associated with symptoms of chronotropic incompetence (B)

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Europace
doi:10.1093/europace/eut082
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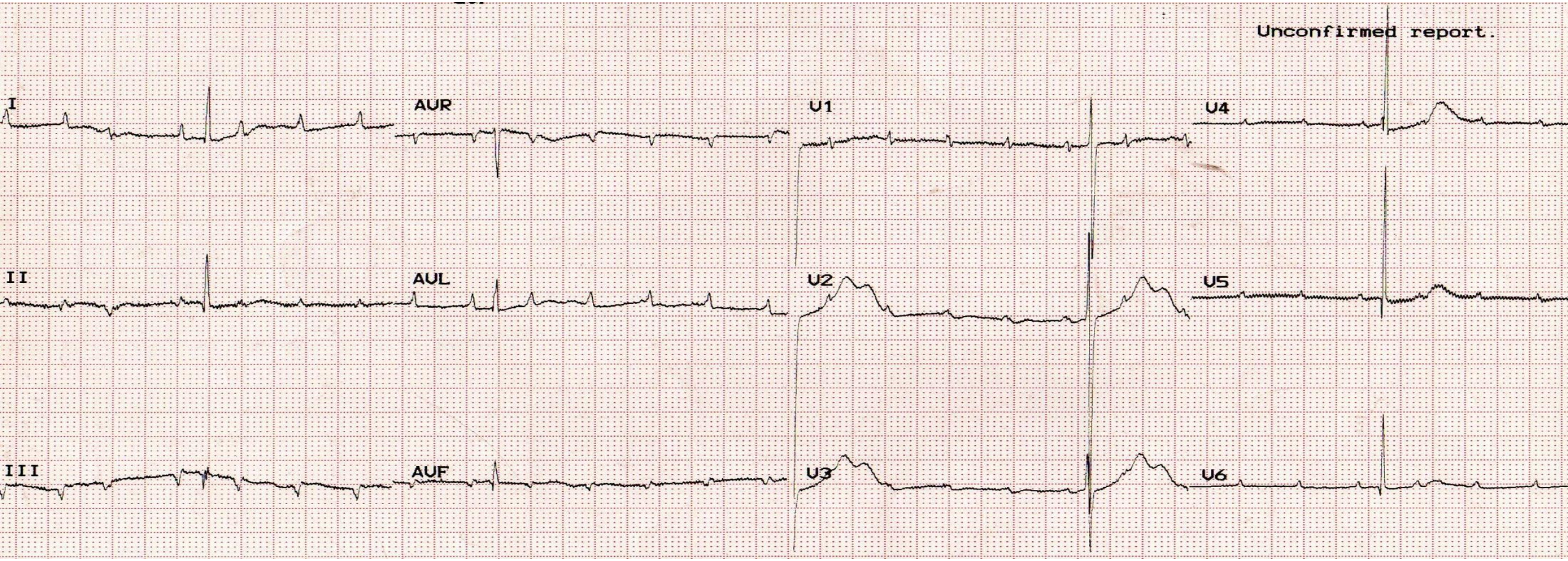
Newborn ECG

Heart rate 35 bpm



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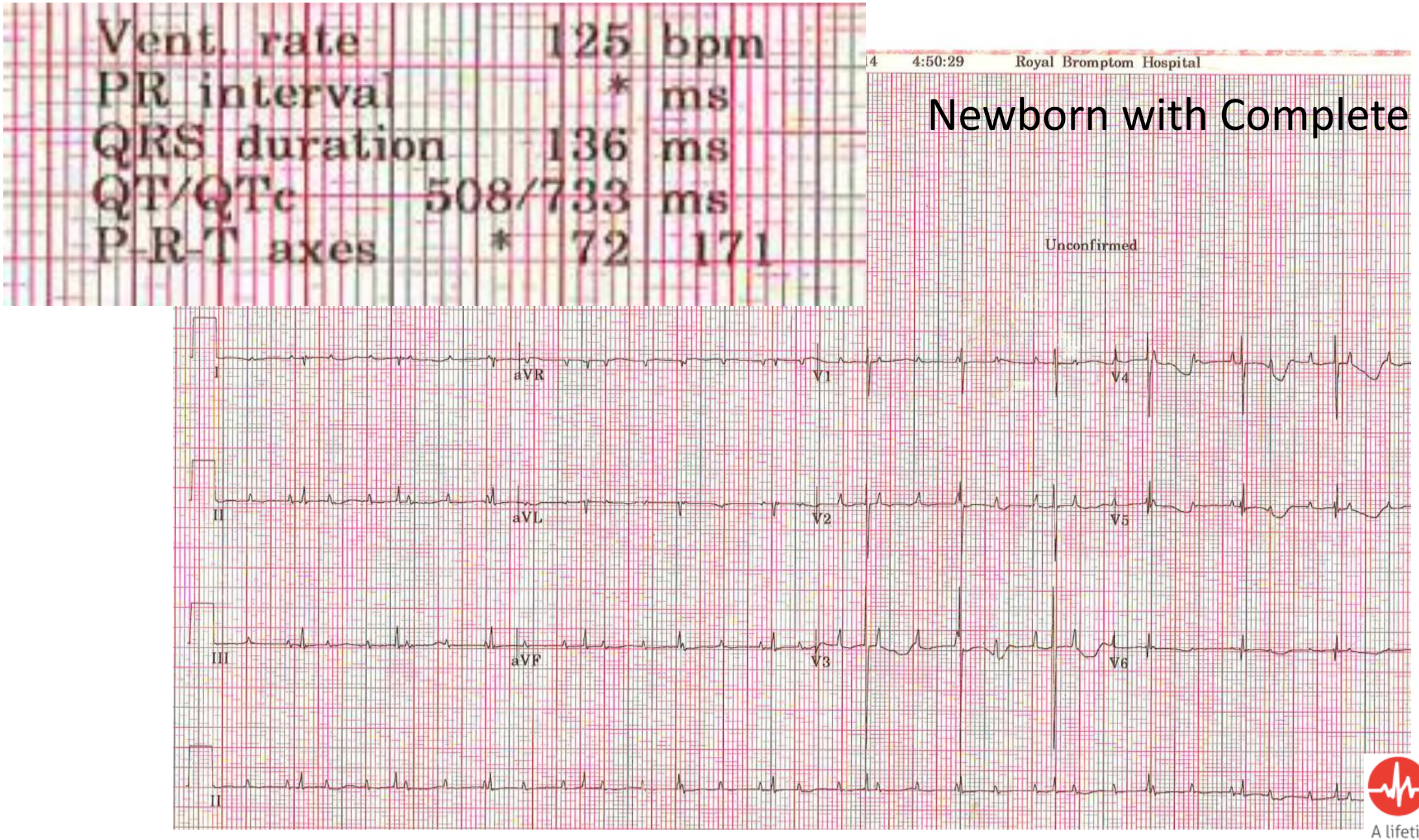
Newborn with Complete AV Block



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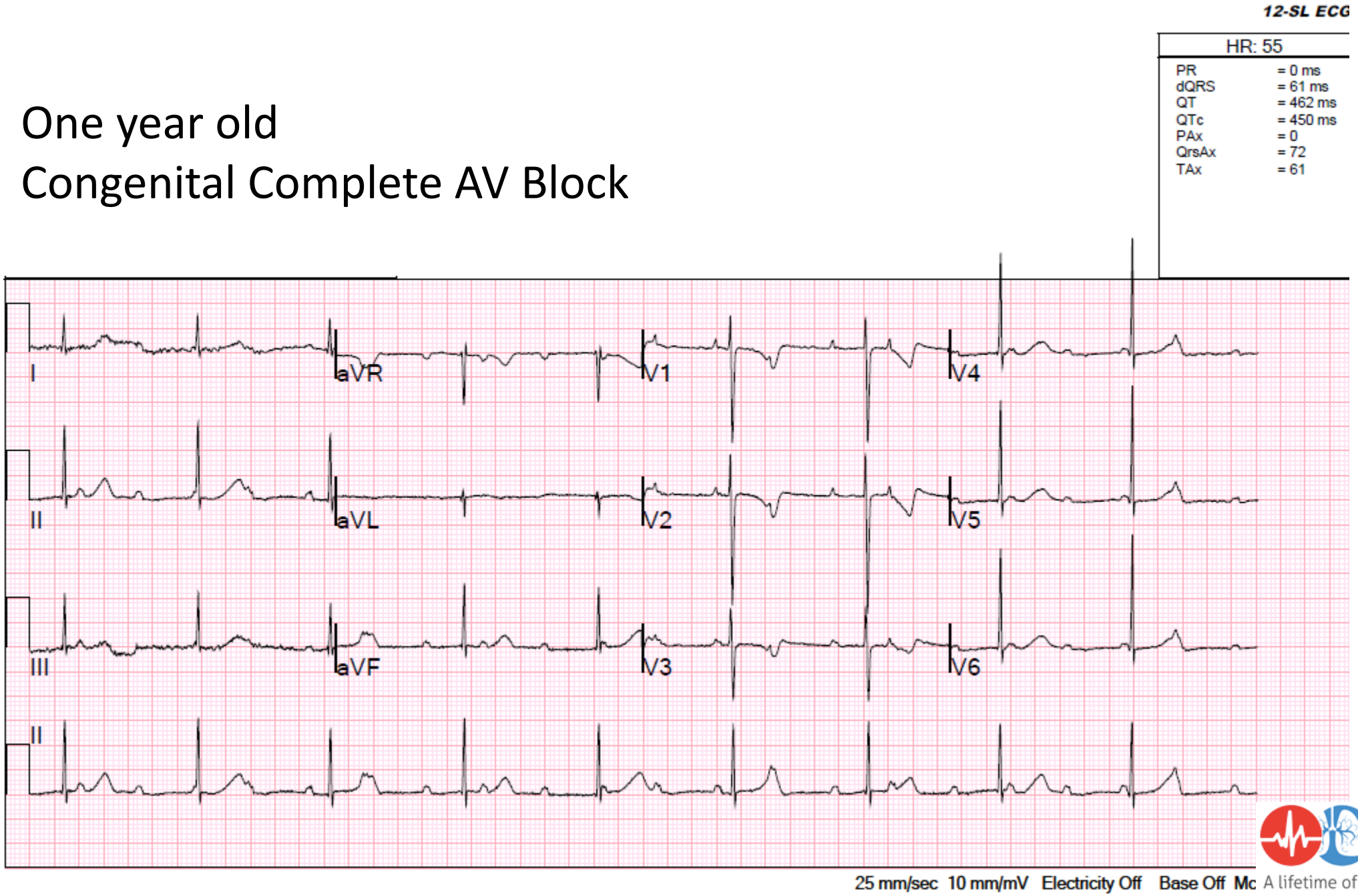
One year old
Congenital Complete AV Block



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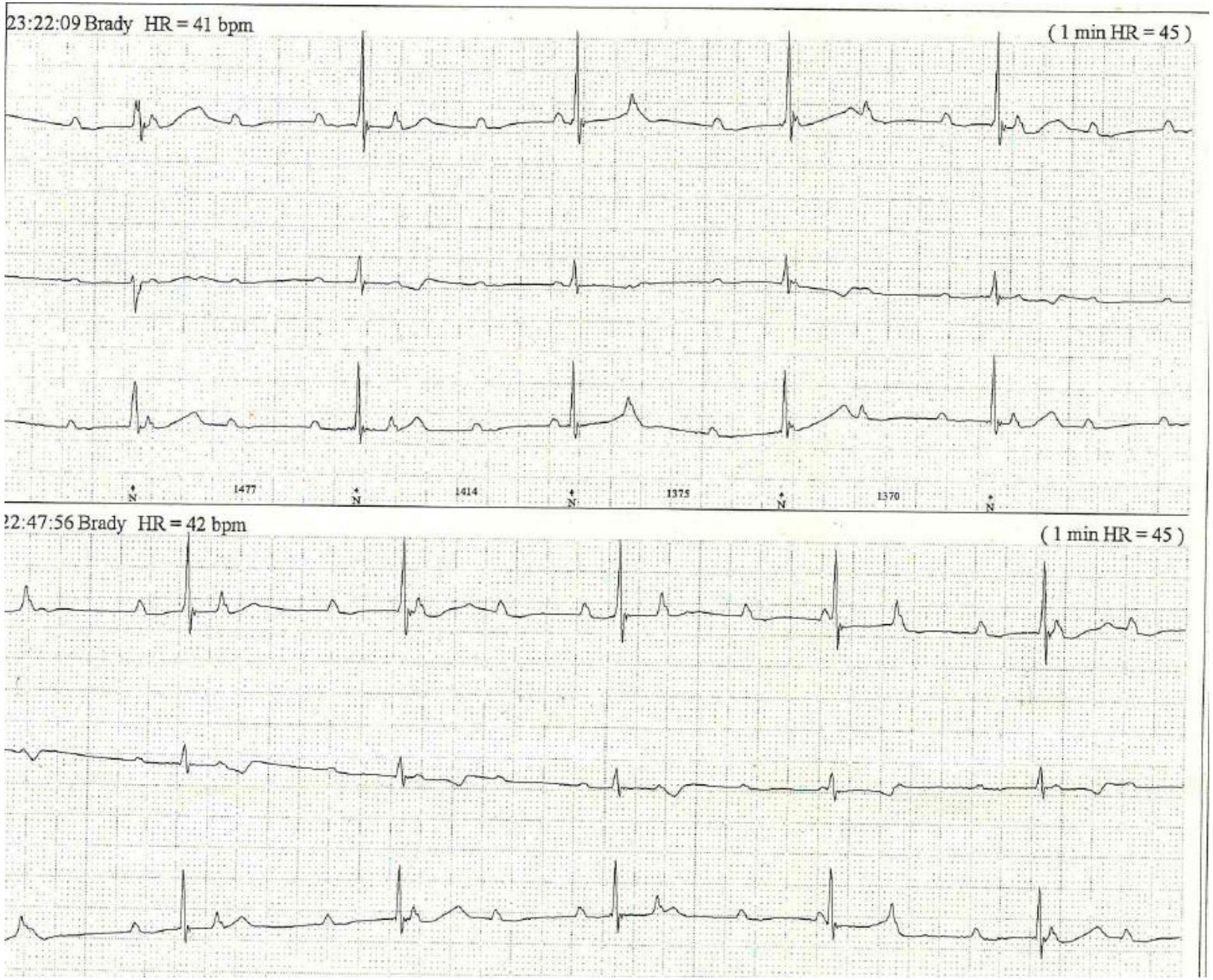

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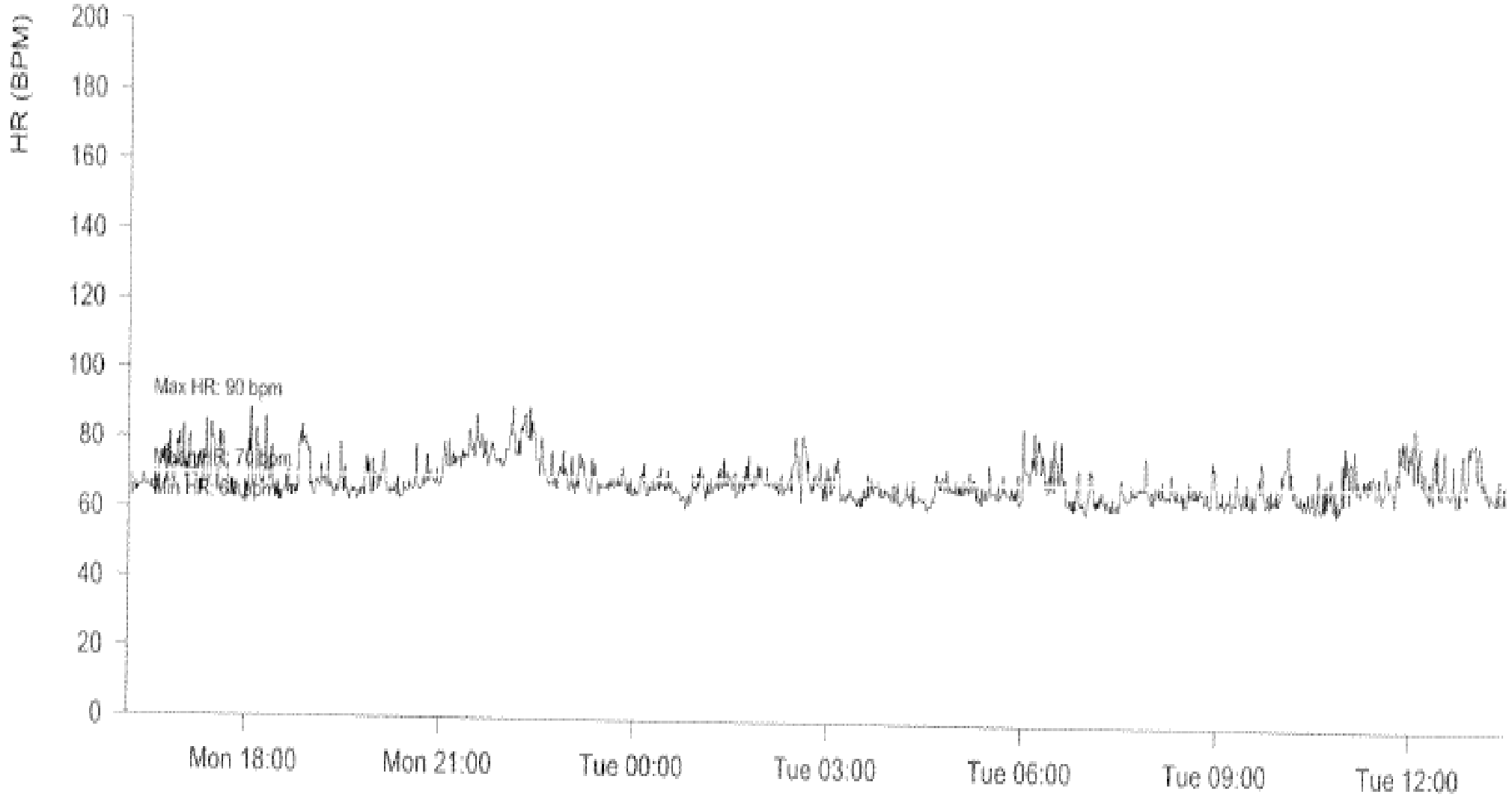

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To pace or not to pace?

Disorders of AV Conduction
Complete congenital atrioventricular block

- Class II
- Complete congenital atrioventricular block in asymptomatic children or adolescents with an acceptable rate, a narrow QRS complex and normal ventricular function (C)

Unconfirmed

Royal Brompton Hospital

26/02/2018 09:48:56 Previous ECG: 22/05/2017 10:02:16

Royal Brompton & Harefield NHS
NHS Foundation Trust

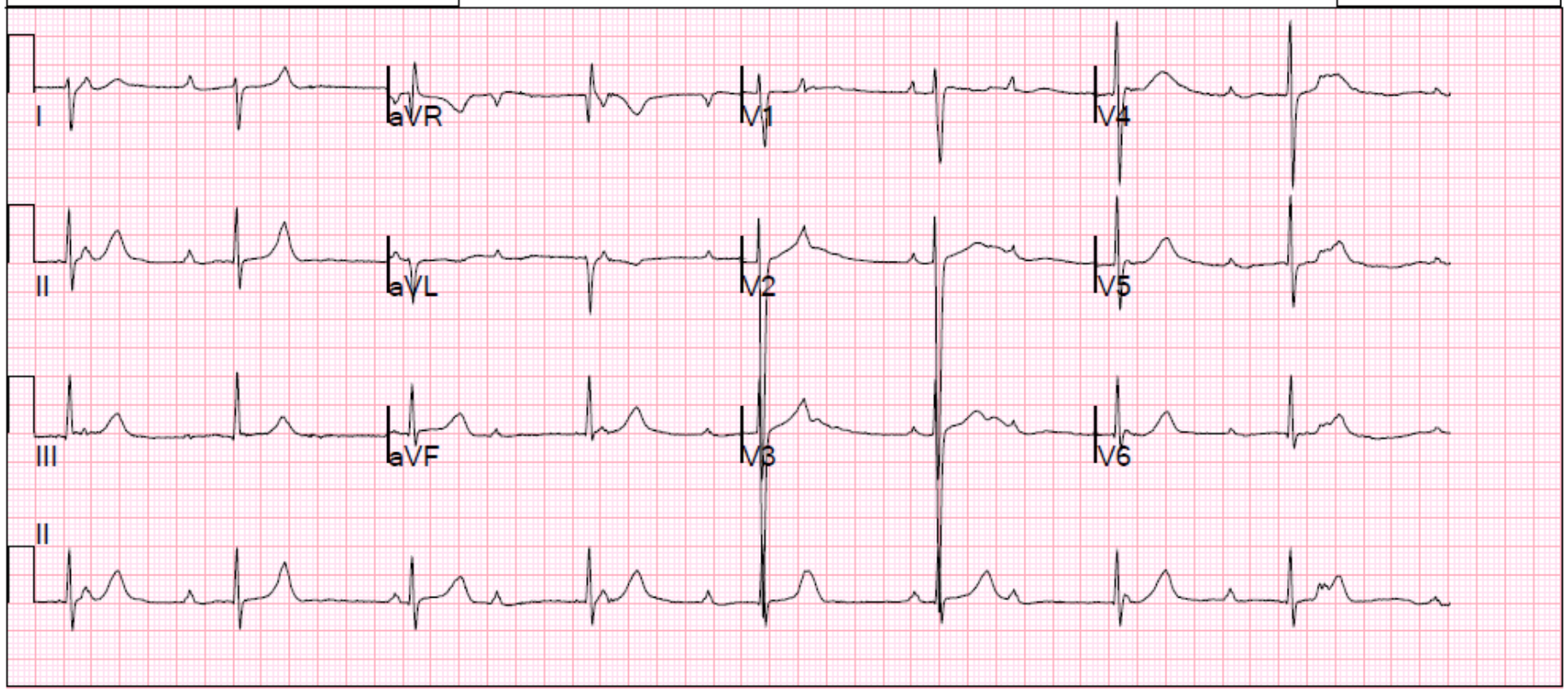
12-SL ECG

Male

12/04/2011 6 year(s)

Referring:
Technician:
Confirmed By:

HR: 48	
PR	= 0 ms
dQRS	= 95 ms
QT	= 479 ms
QTc	= 446 ms
PAx	= 0
QrsAx	= 124
TAx	= 70



25 mm/sec 10 mm/mV F: -1 Hz W: -0.01--1 Hz Mckesson - MIG

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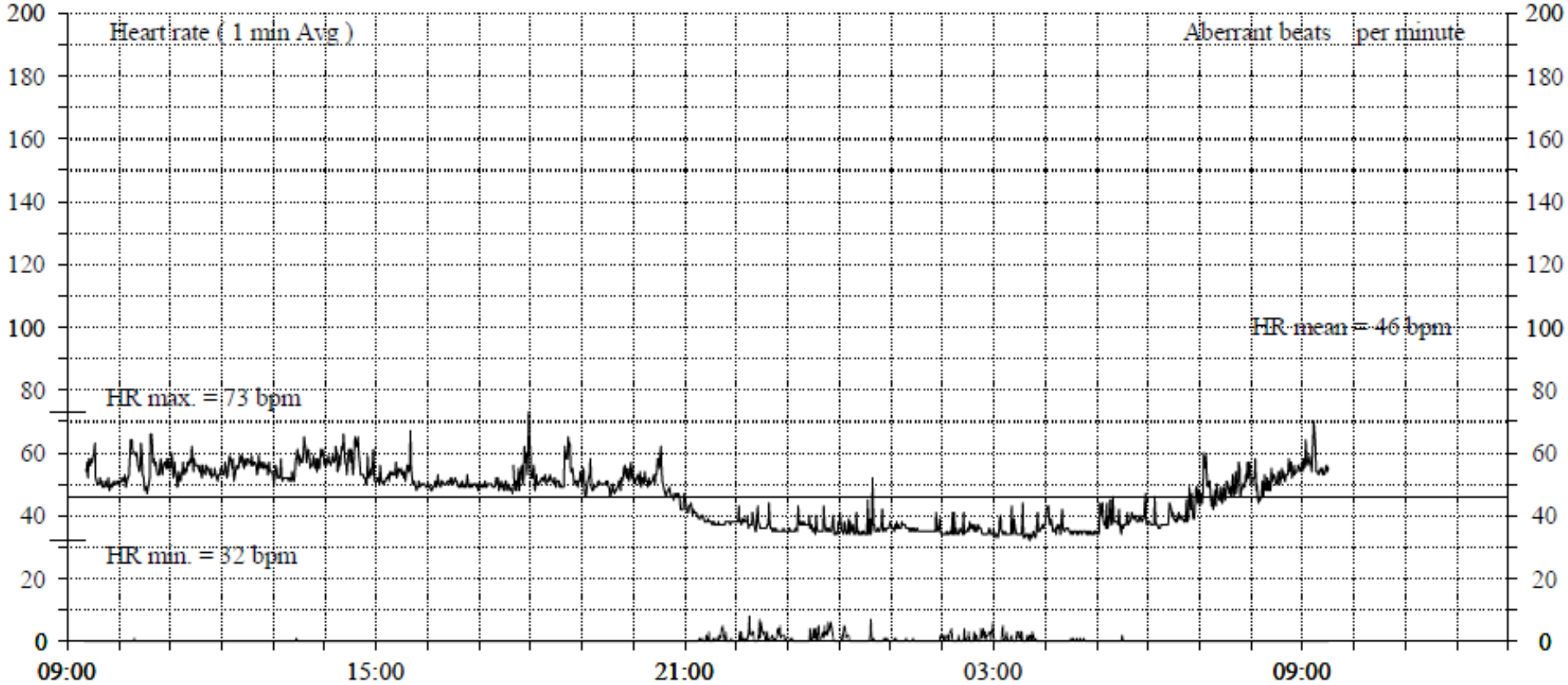

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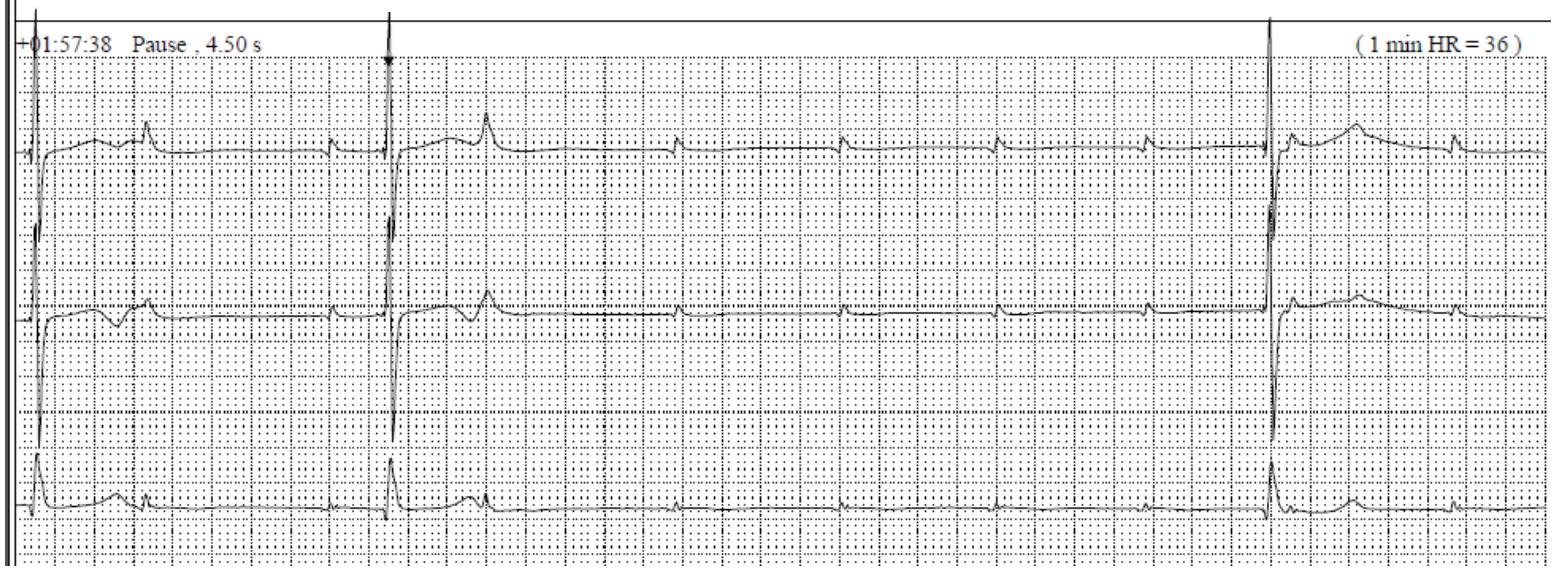
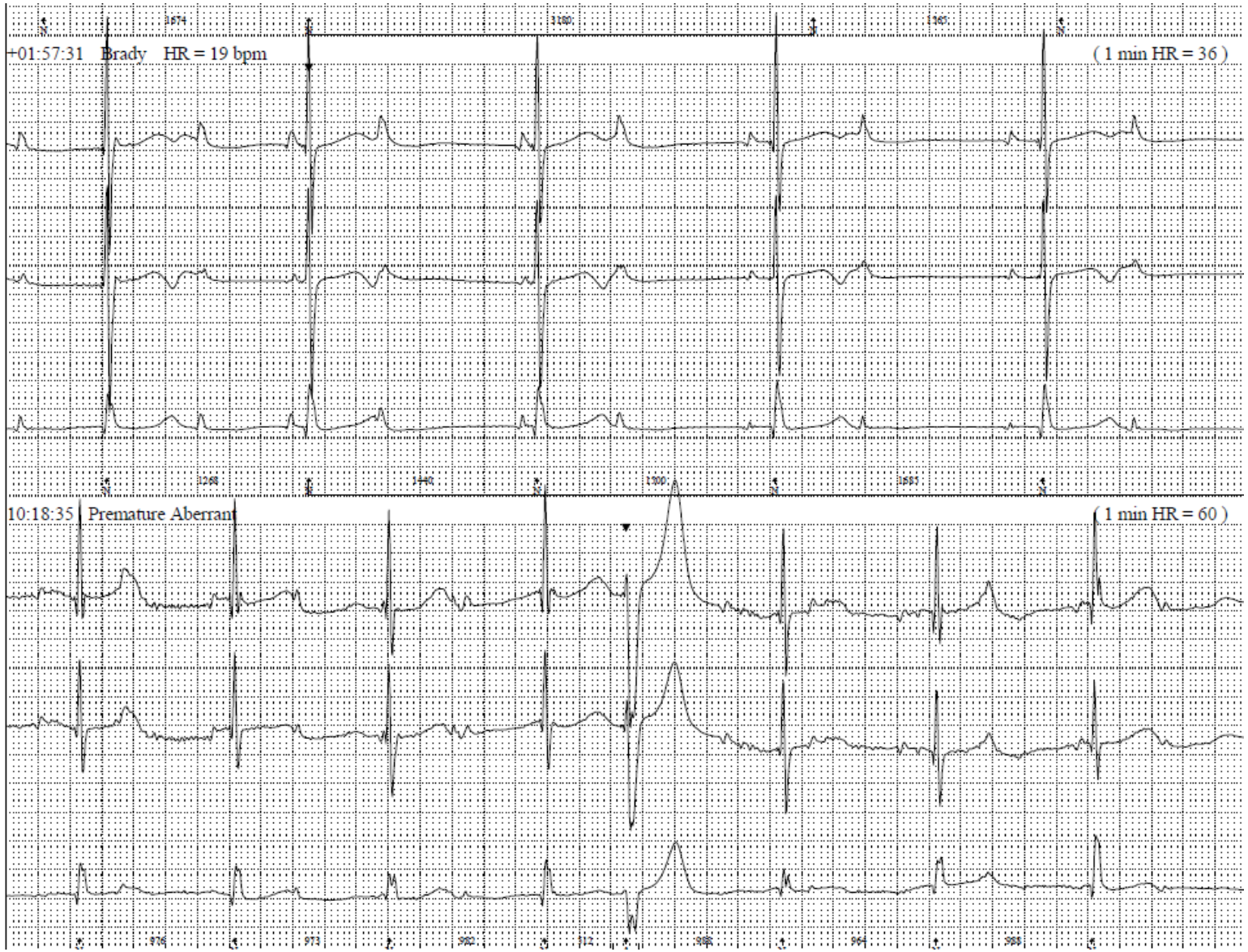
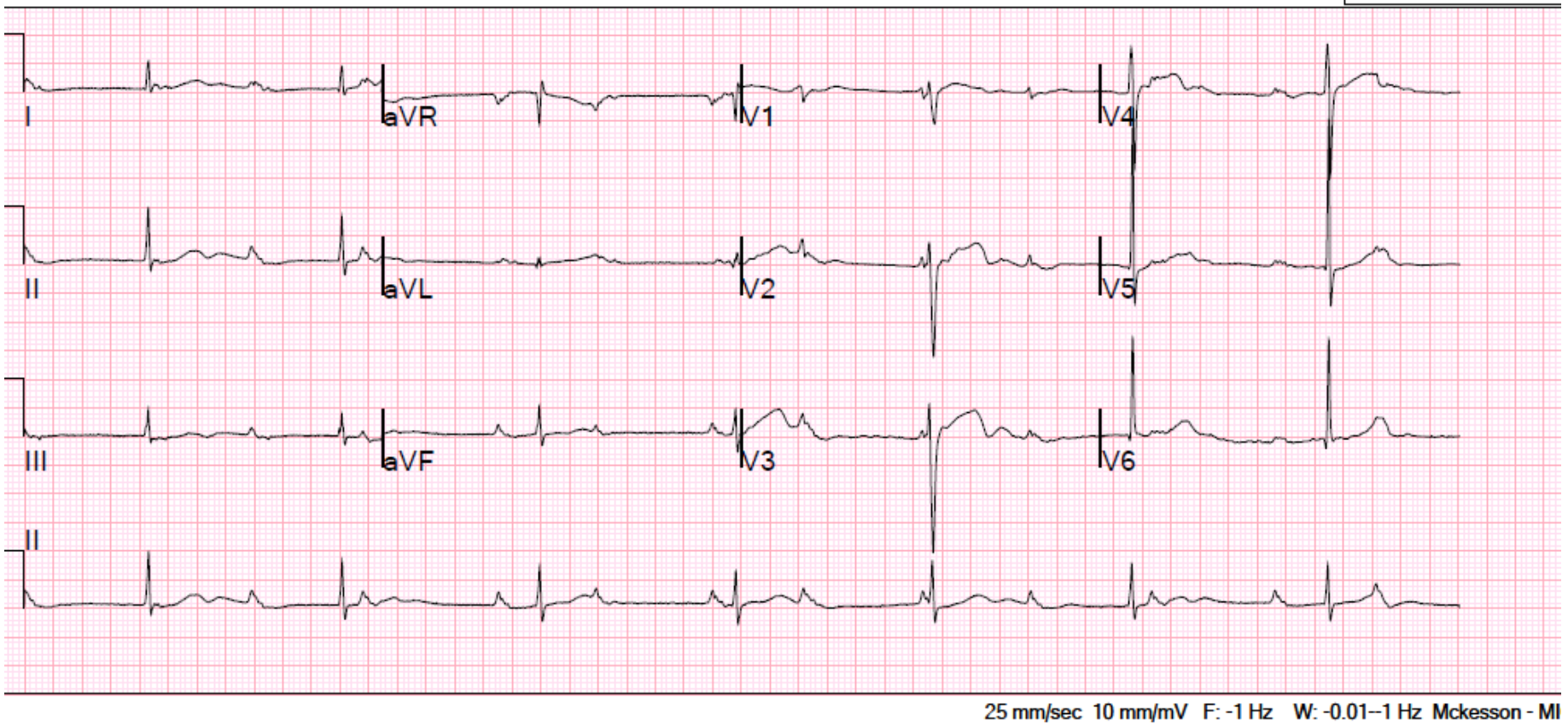
Recording Date : 04/09/2017




+01:57:31 Brady HR = 19 bpm
(1 min HR = 36)
10:18:35 Premature Aberrant
(1 min HR = 60)

Asymptomatic
10 year old
Congenital AV Block



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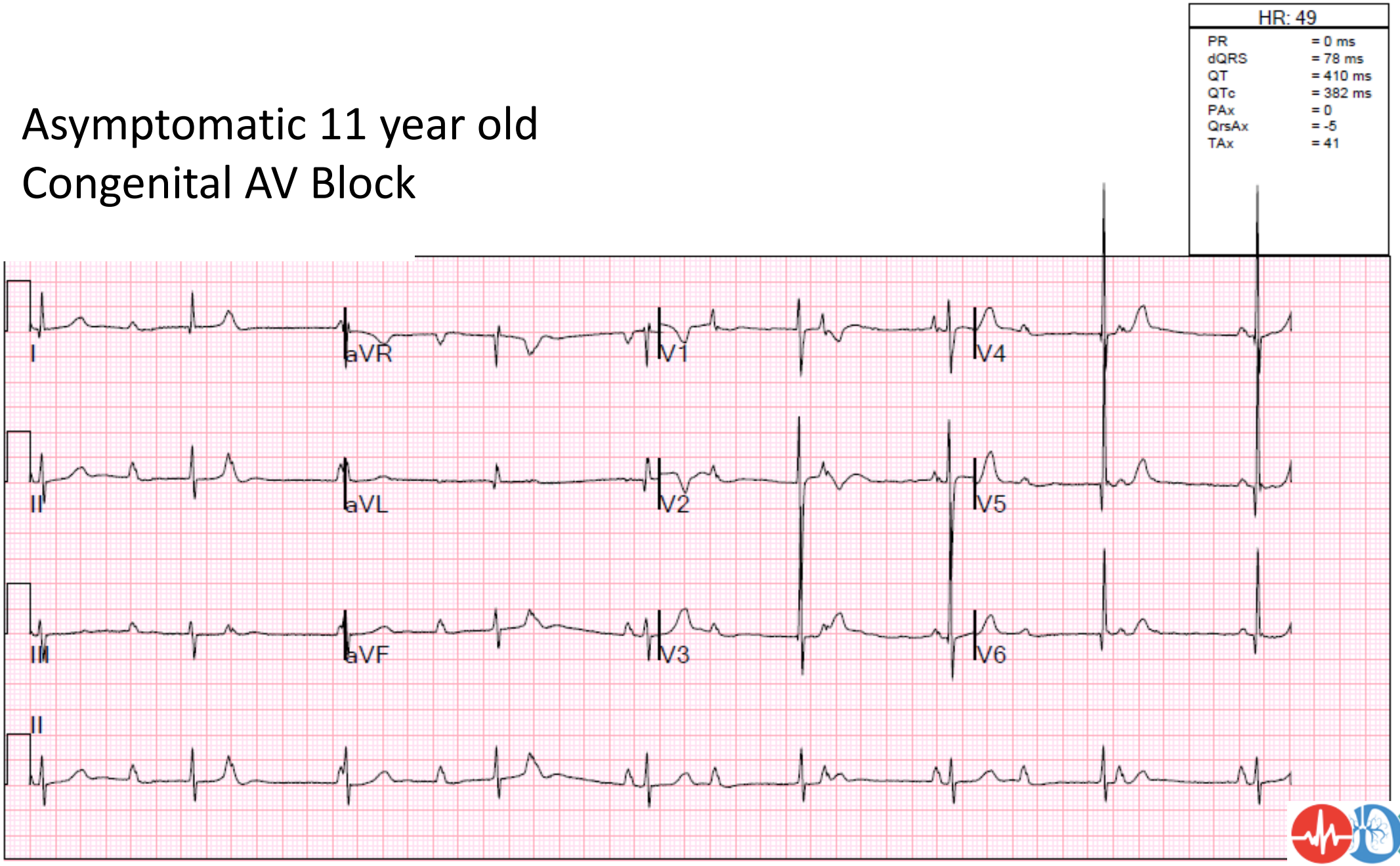
Asymptomatic 11 year old
Congenital AV Block



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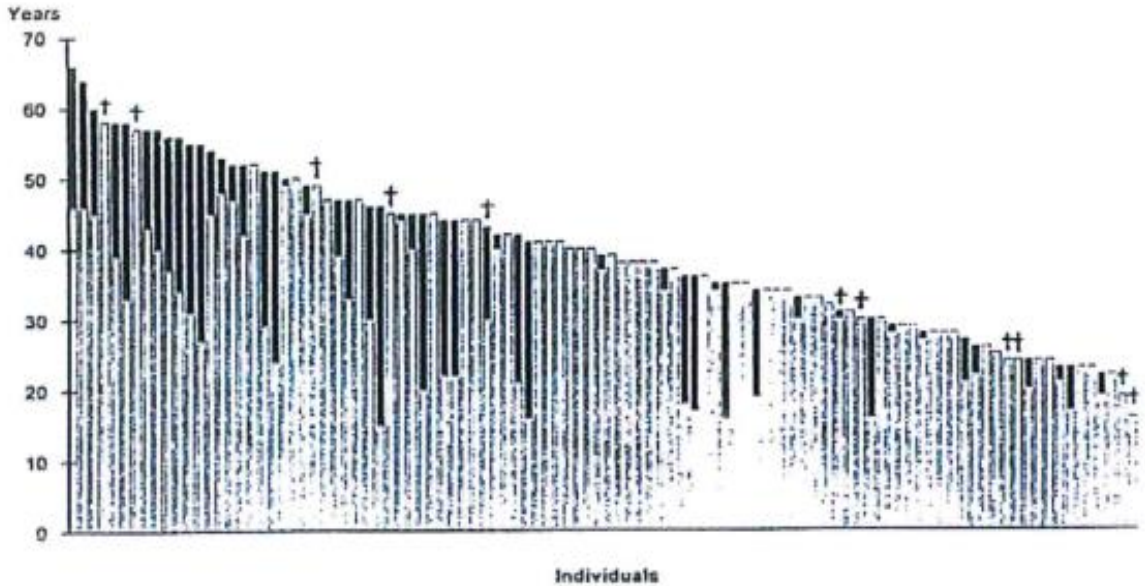
Isolated Congenital Complete Atrioventricular Block in Adult Life

A Prospective Study

Magnus Michaëlsson, MD, PhD; Anders Jonzon, MD, PhD; Tomas Riesenfeld, MD, PhD

Circulation 1995



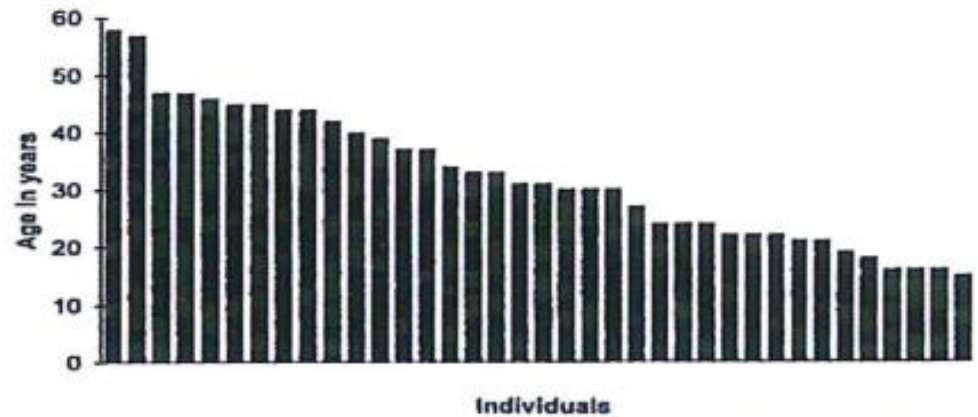
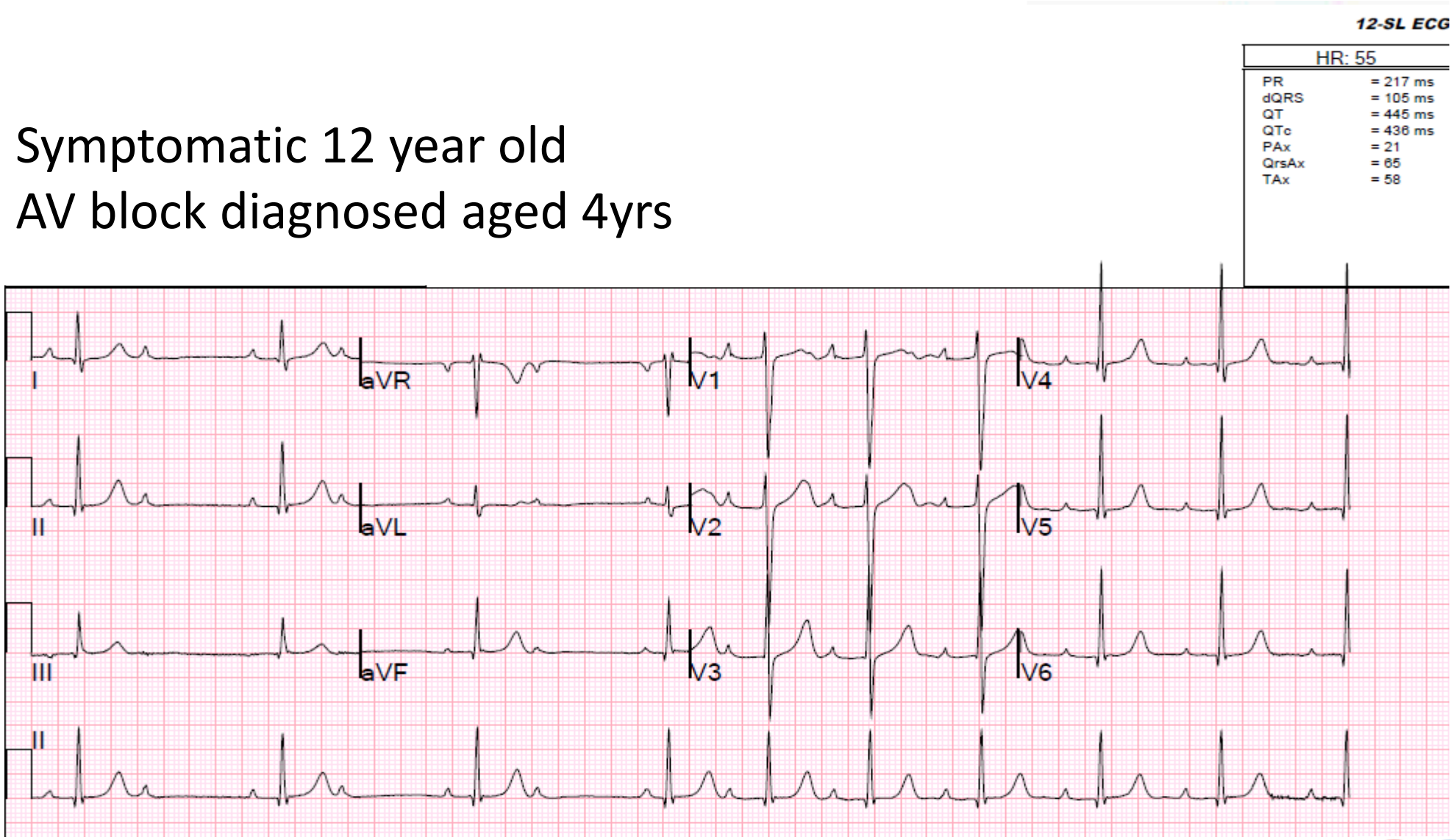


FIG 2. Bar graph showing the age at death, syncope, or presyncope in 37 patients.

Symptomatic 12 year old
AV block diagnosed aged 4yrs



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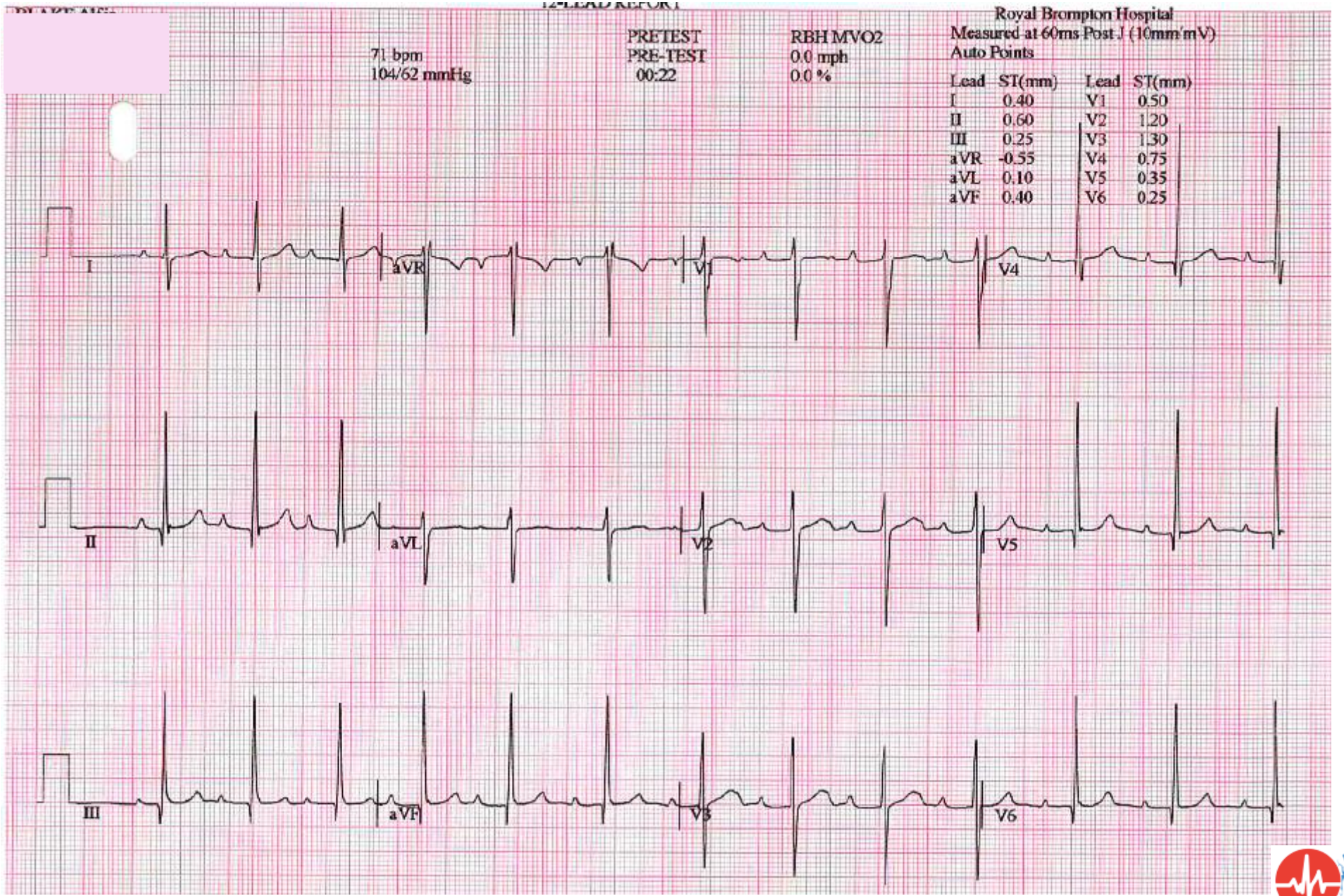
Non surgical atrioventricular block

- Class 1
  - Advanced second or third degree AV block associated with symptomatic bradycardia, ventricular dysfunction or low cardiac output (C)



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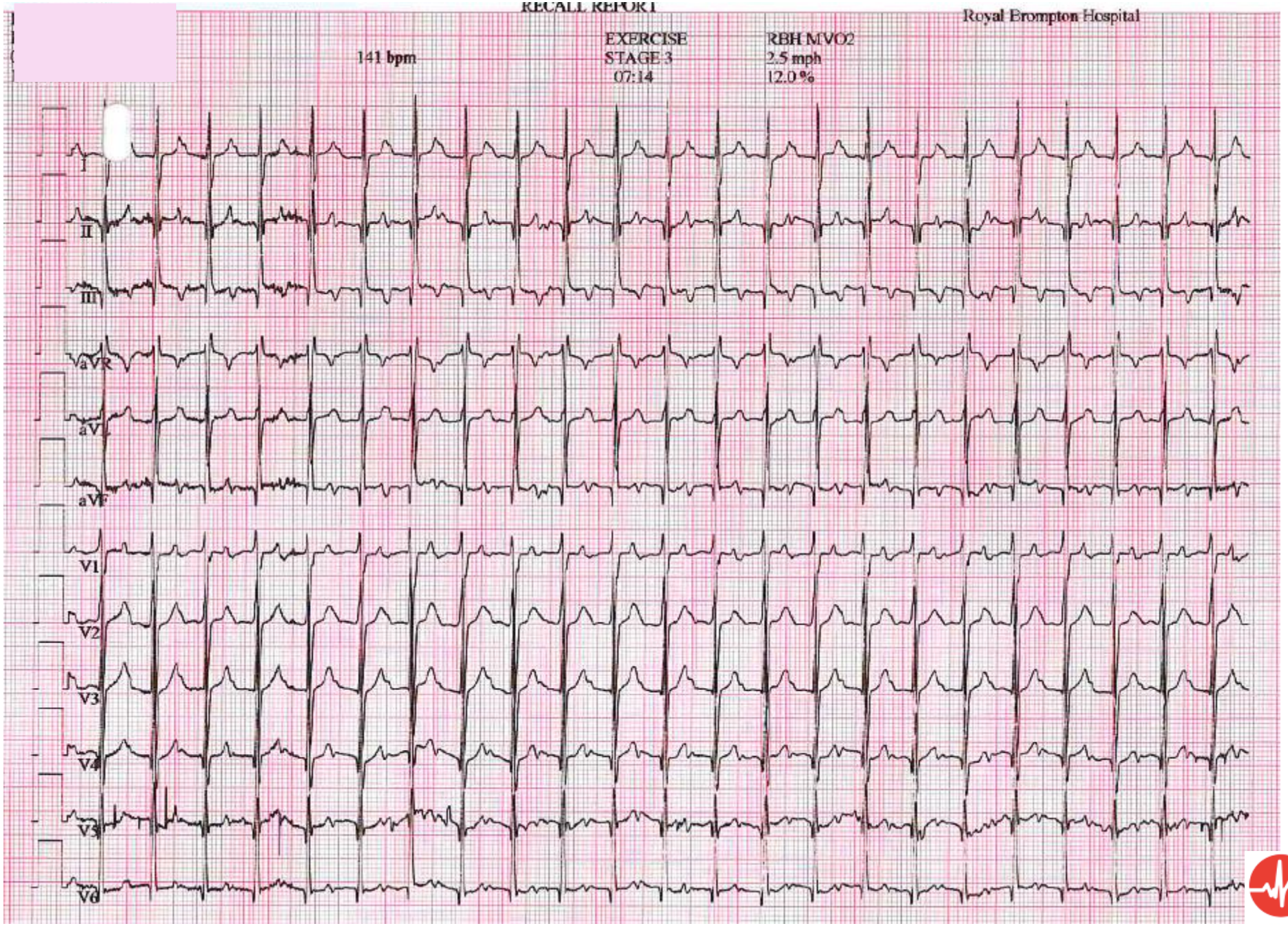

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Post surgery

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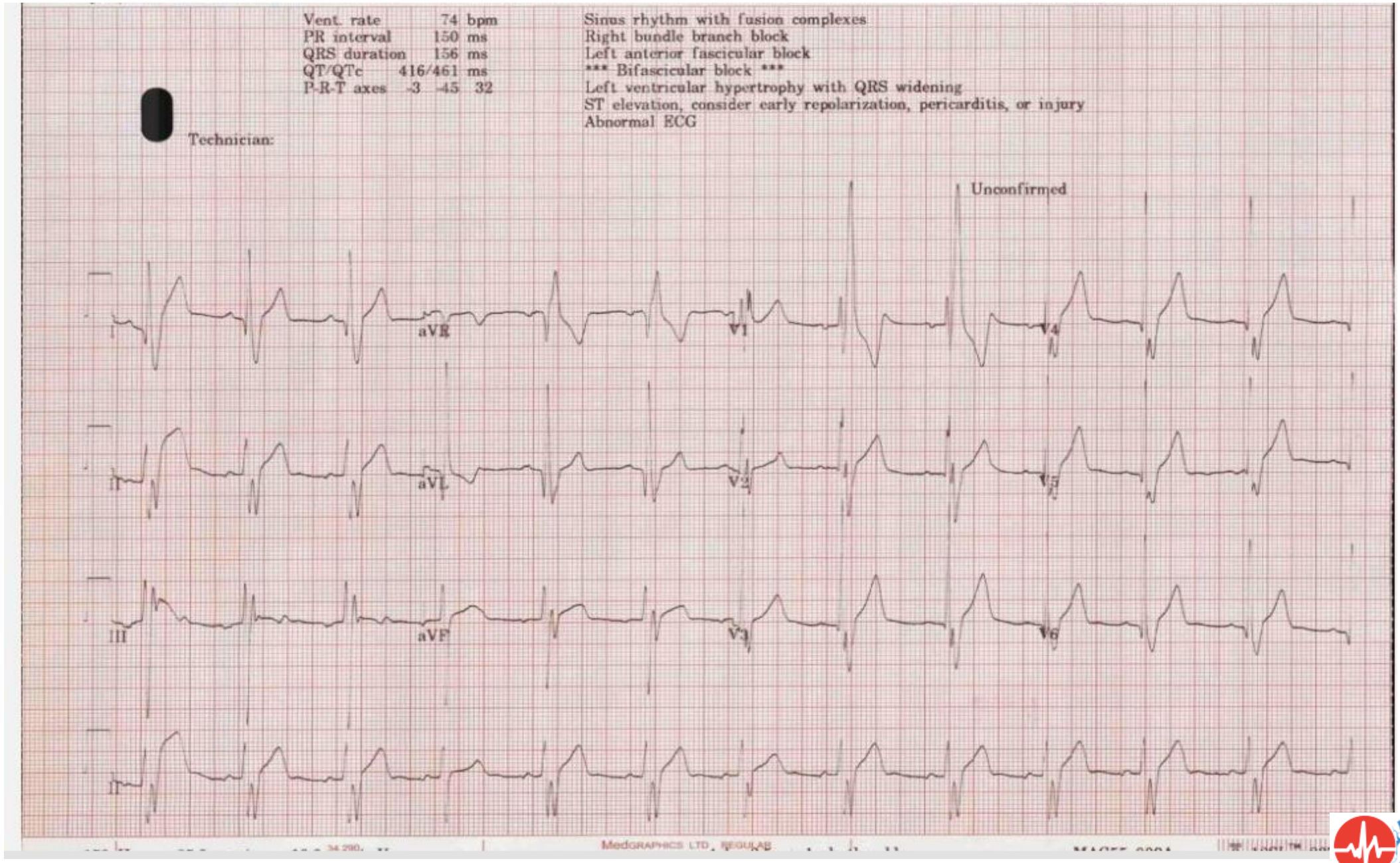
Post operative atrioventricular block

- Class 1
  - Post operative advanced second or third degree AV block not expected to resolve or persisting at least 7 days after cardiac surgery (B)
- Class IIb
  - Transient post operative third degree AV block with residual bifascicular block (C)

Europace
doi:10.1093/europace/eut082



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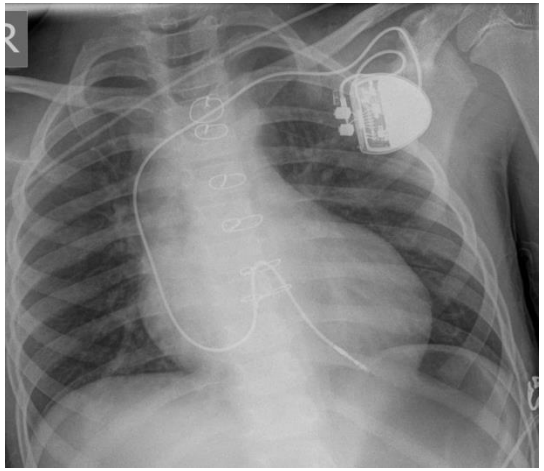
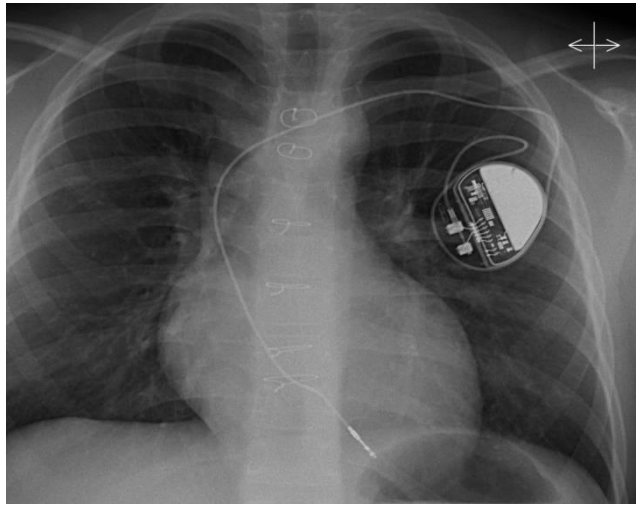
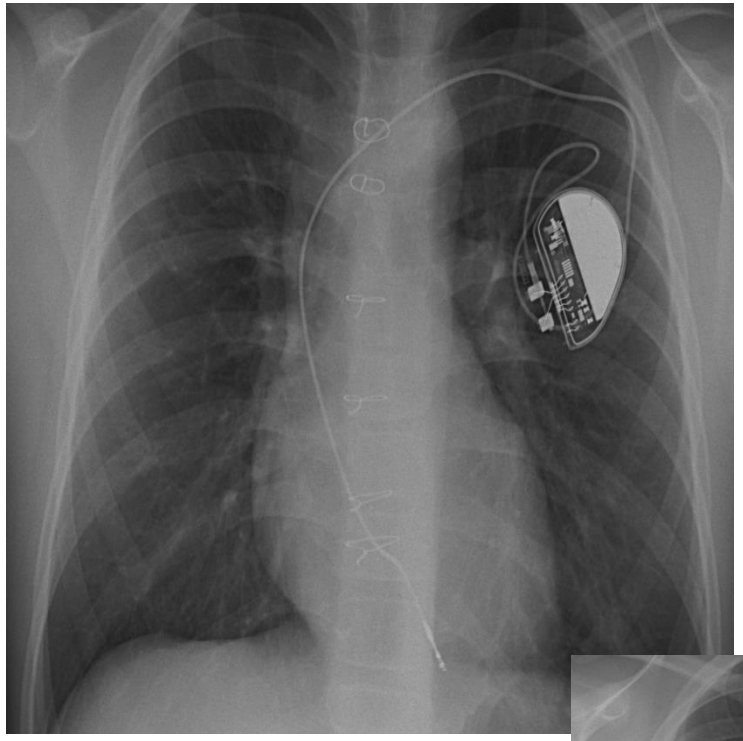


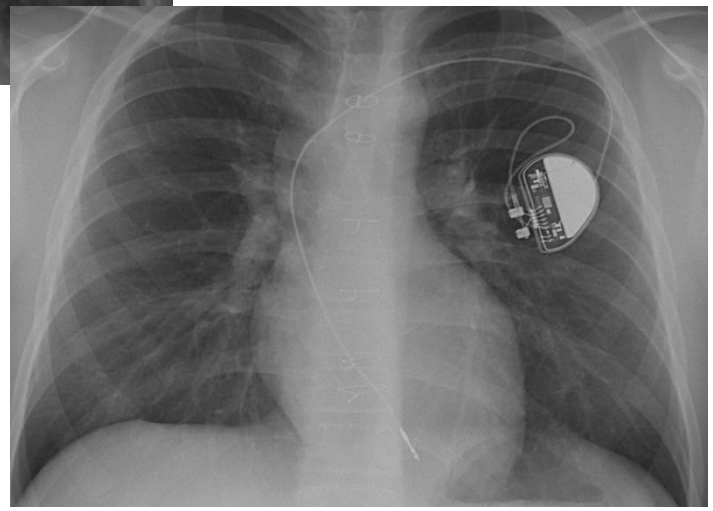
Implant post surgery
Infant



Age 8 years



Age 10 years



12 years

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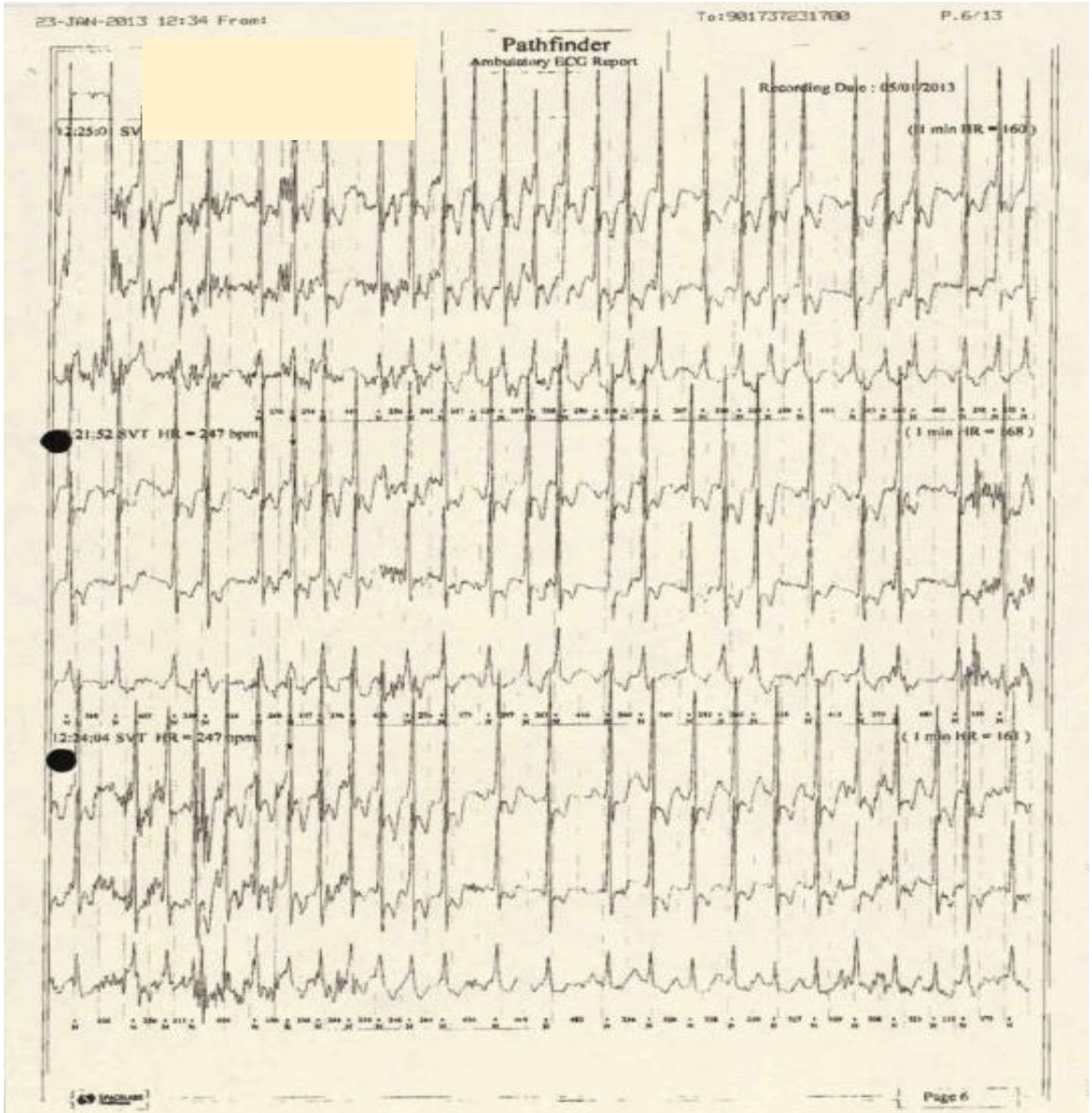
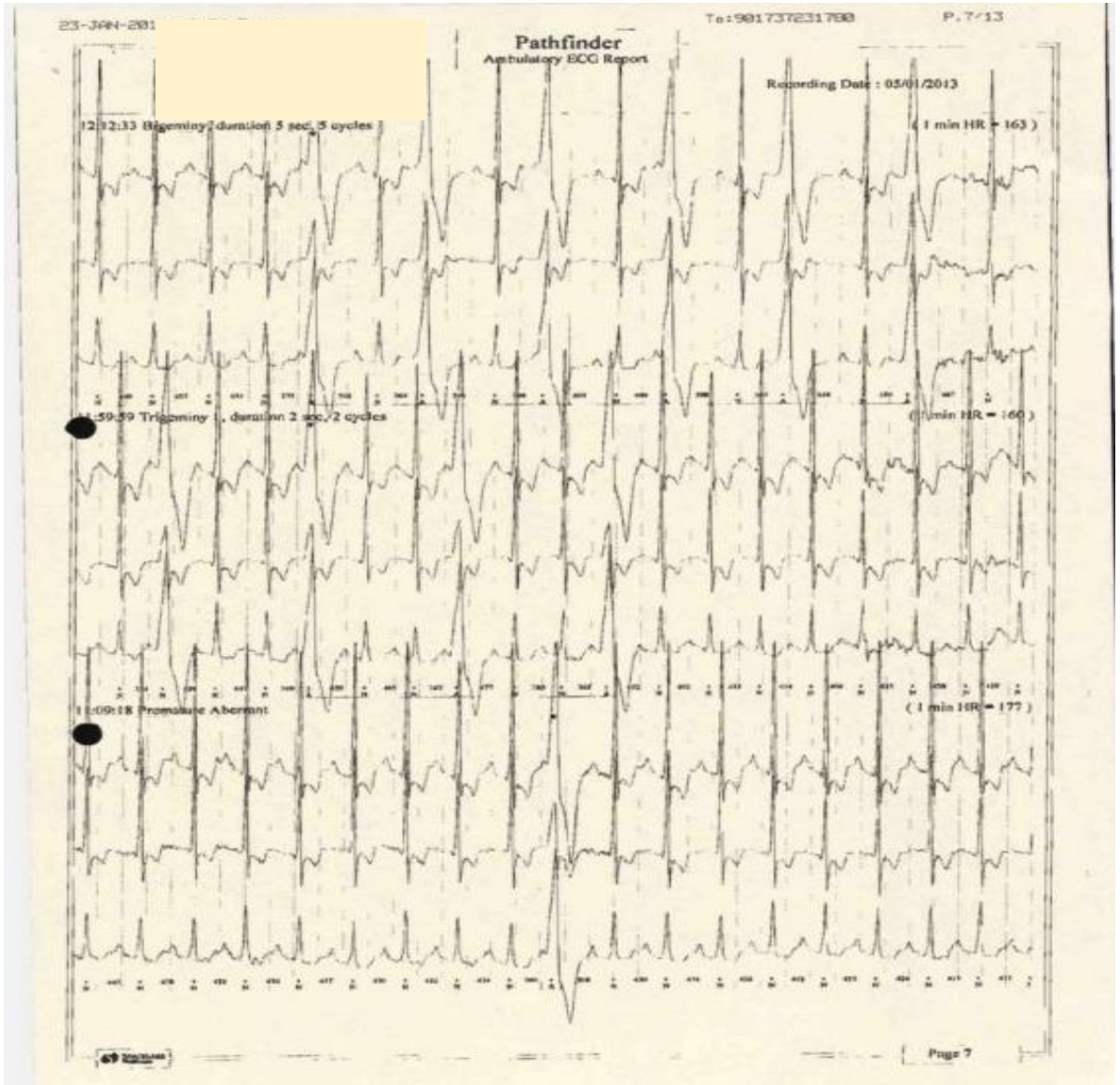
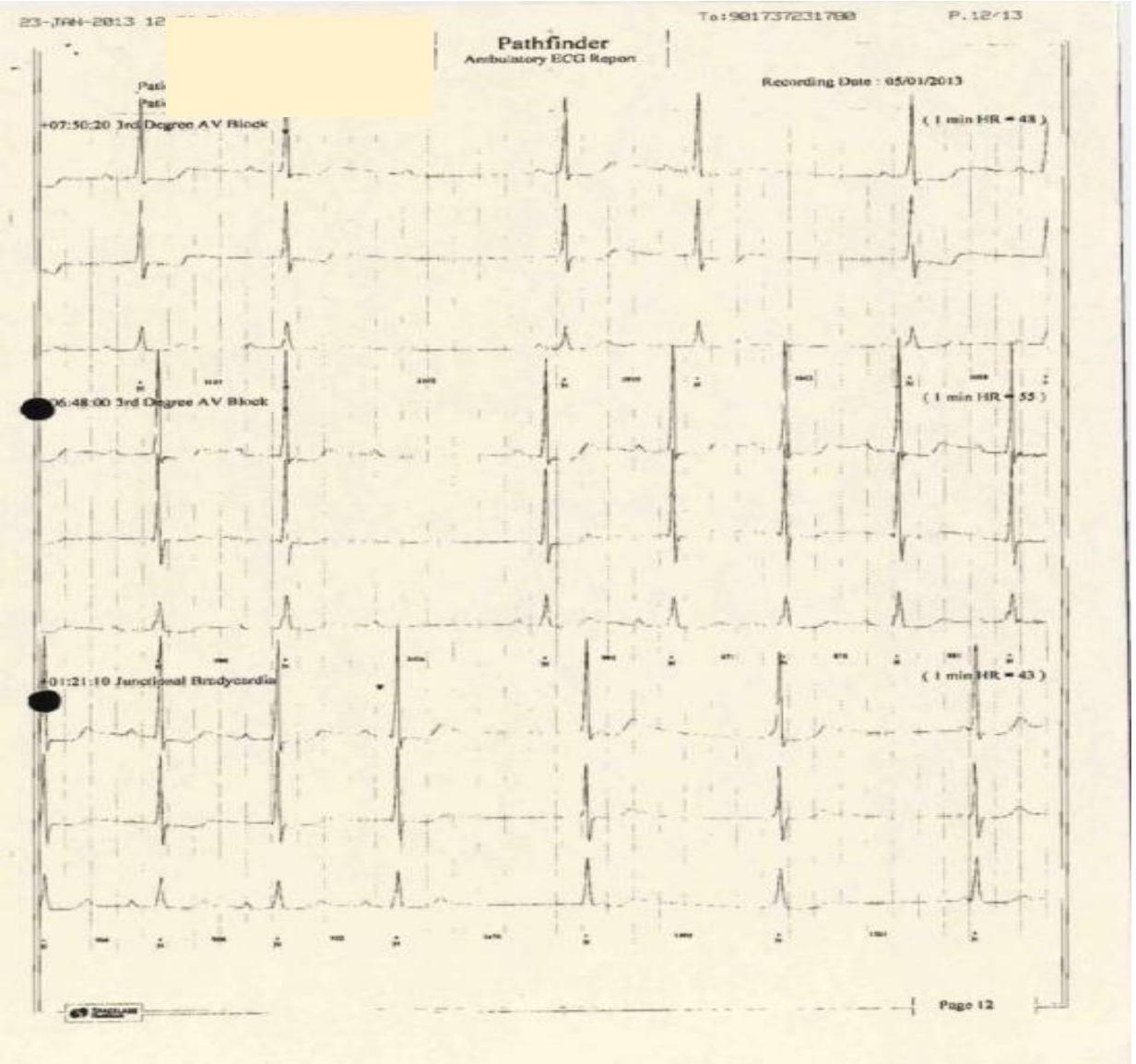
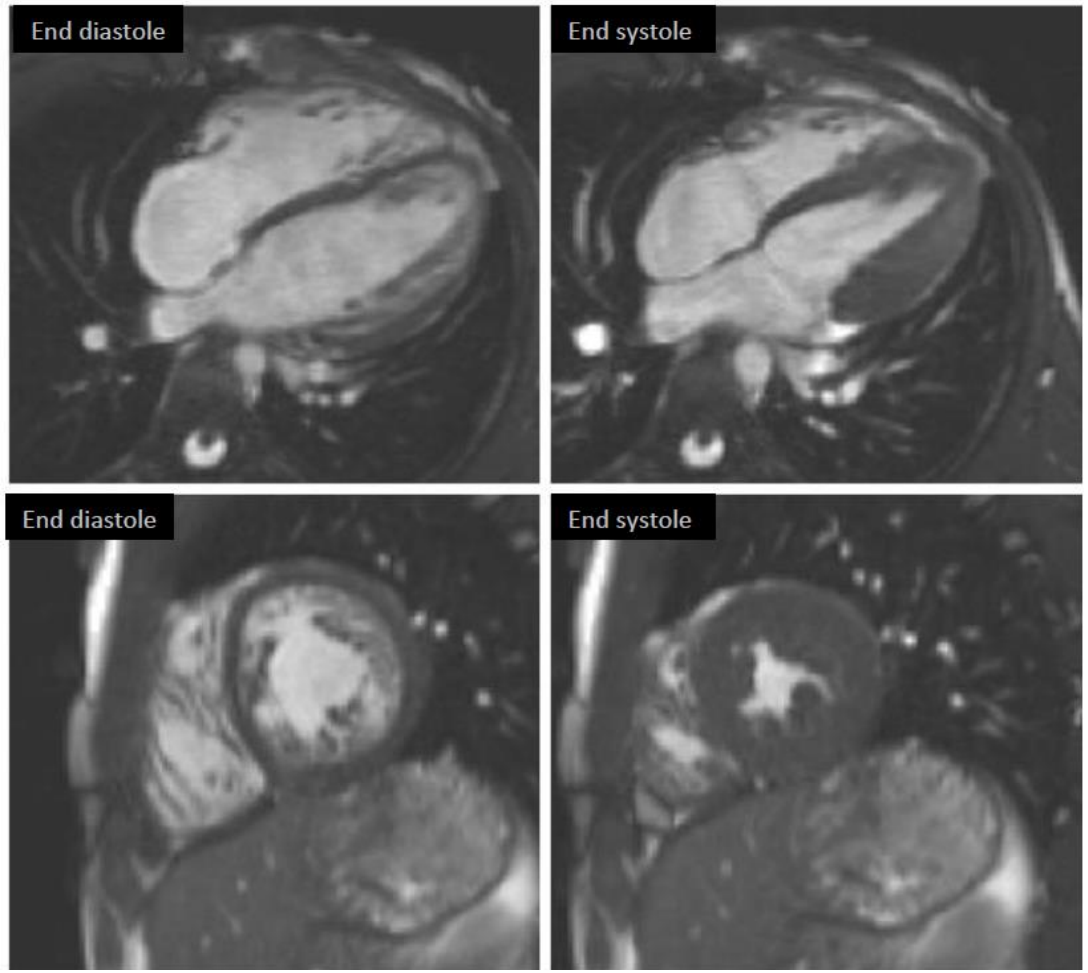
Others:
Neuromuscular disease associated with AV conduction disease eg myotonic muscular dystrophy, Kearns Sayre syndrome, Erb dystrophy (limb girdle) peroneal muscular atrophy

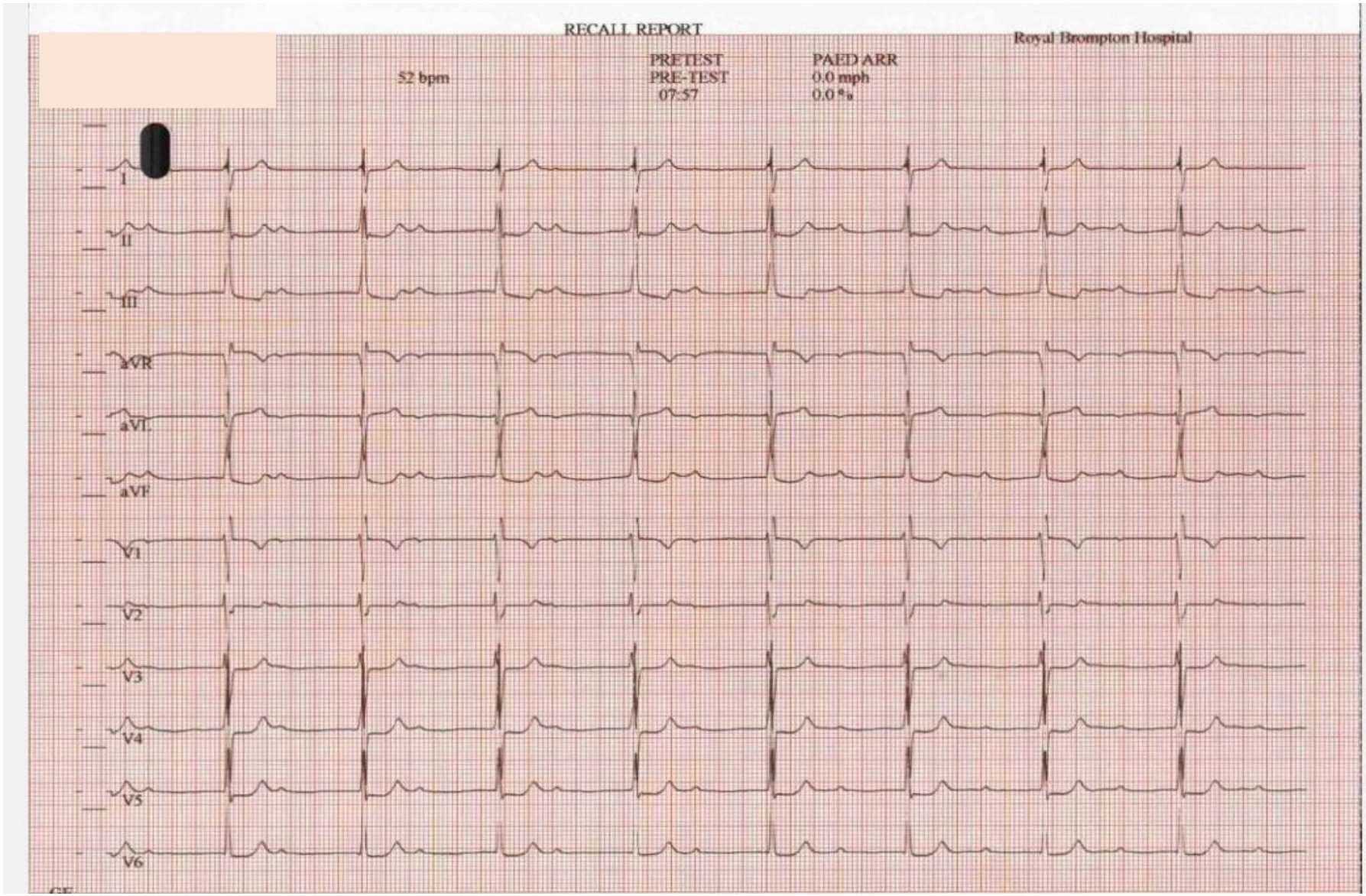
- Class 1
- Third degree or advanced second degree AV block with or without symptoms (B)
- Class IIb
- Any degree of AV block, because the progression of the conduction disease may be unpredictable (B)


23-JAN-2013 12:34 From:
To:901737231700
P.6/13
Pathfinder
Ambulatory ECG Report
Recording Date : 05/01/2013
(1 min HR = 163)
12:25:0 SV
21:52 SVT HR = 247 bpm
(1 min HR = 168)
12:24:04 SVT HR = 247 bpm
(1 min HR = 163)
Page 6


23-JAN-201
To:901737231700
P.7/13
Pathfinder
Ambulatory ECG Report
Recording Date : 05/01/2013
12:32:33 Bigeminy, duration 5 sec, 5 cycles
(1 min HR = 163)
11:59:59 Trigeminy , duration 2 sec, 2 cycles
(1 min HR = 160)
11:09:18 Premature Aberrant
(1 min HR = 177)
Page 7


23-JAN-2013 12
To:9017372317
P.12/13
Pathfinder
Ambulatory ECG Report
Recording Date : 05/01/2013
07:50:20 3rd Degree AV Block
(1 min HR = 48)
06:48:00 3rd Degree AV Block
(1 min HR = 55)
01:21:10 Junctional Bradycardia
(1 min HR = 43)
Page 12


End diastole
End systole
End diastole
End systole


RECALL REPORT
Royal Brompton Hospital
52 bpm
PRETEST
PRE-TEST
07:57
PAED ARR
0.0 mph
0.0 %
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

Non-sense mutation in NKX 2-5

Unconfirmed

Royal Brompton Hospital

06/03/2018 16:01:24

Previous ECG: 29/11/2017 09:49:16

Royal Brompton & Harefield NHS
NHS Foundation Trust

12-SL ECG

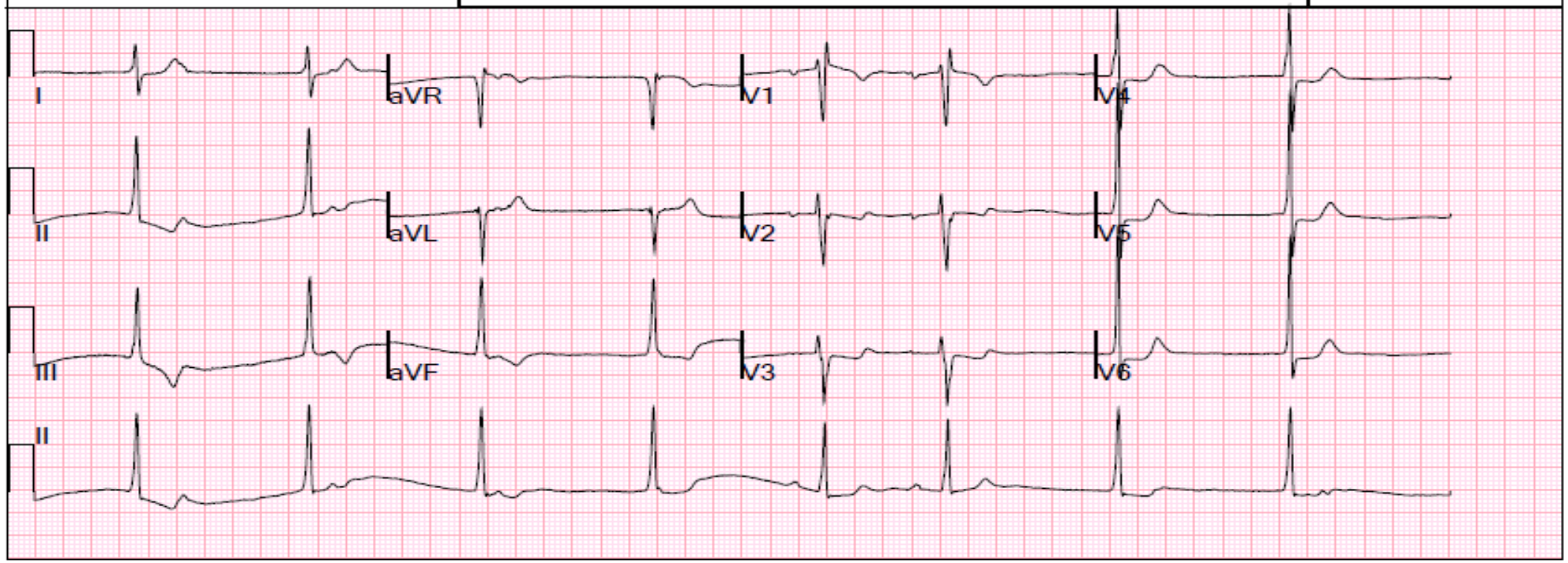
Male

Comments:

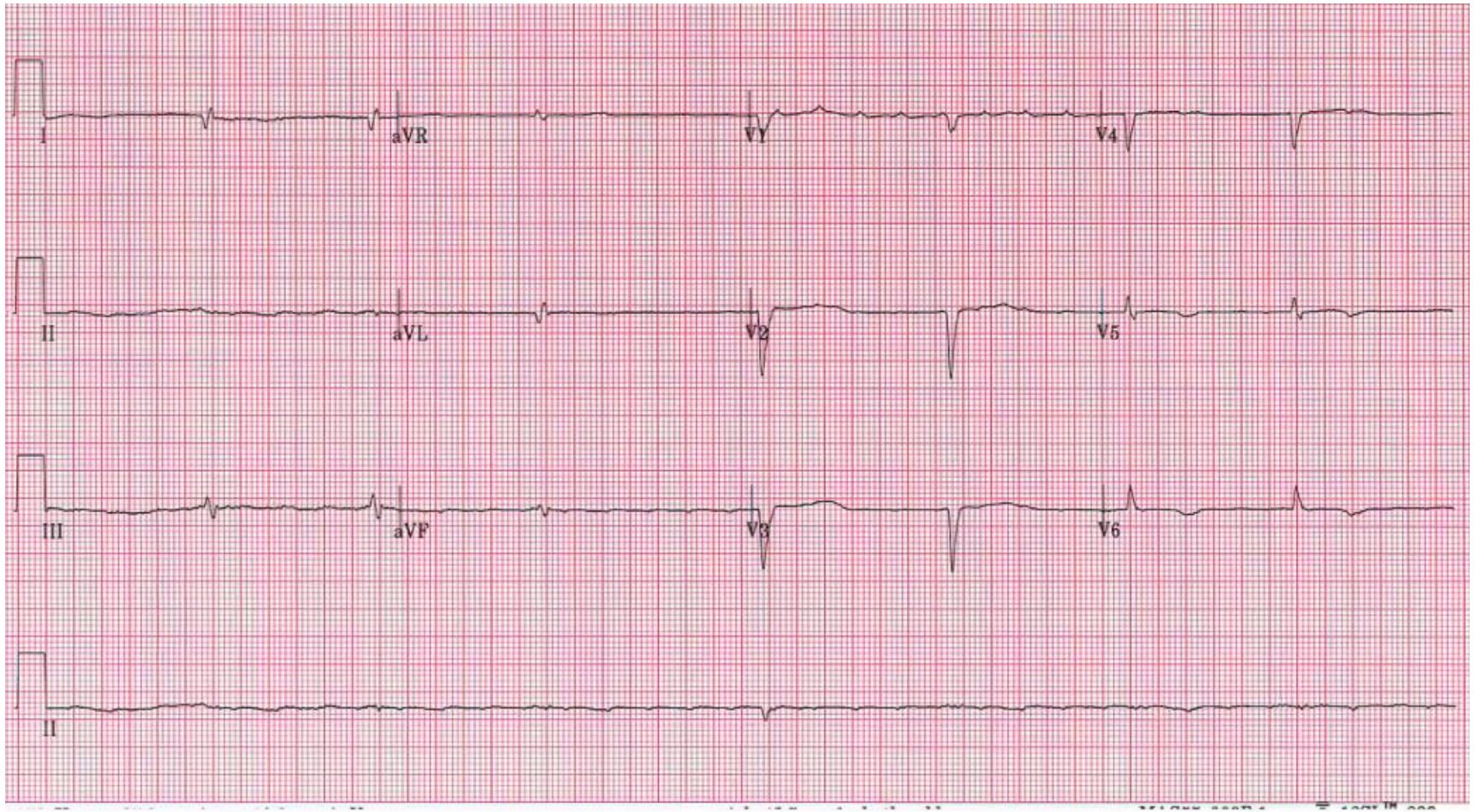
HR: 51

PR	= 0 ms
dQRS	= 102 ms
QT	= 436 ms
QTc	= 413 ms
PAx	= 0
QrsAx	= 88
TAx	= -22

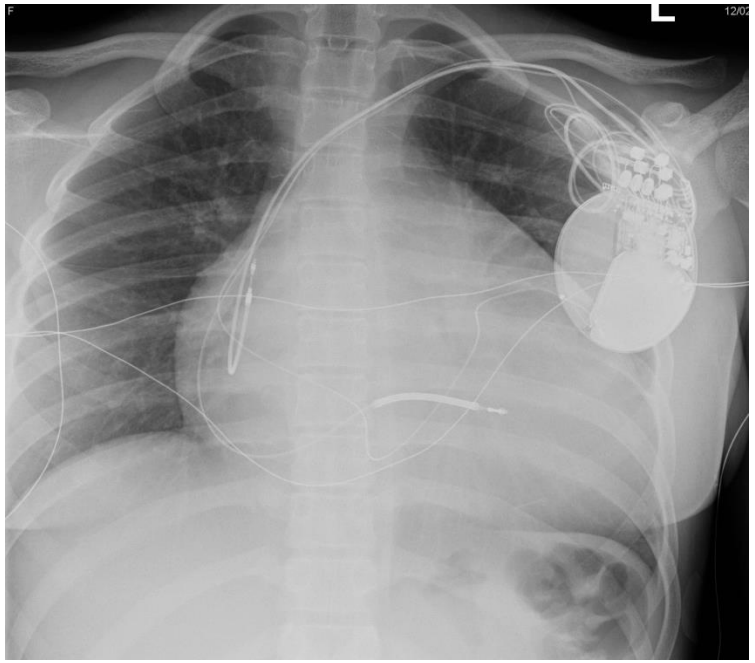
Referring:
Technician:
Confirmed By:



25 mm/sec 10 mm/mV F: -1 Hz W: -0.01--1 Hz Mckesson - MIG



Page 1 of 1 PICU



Aetiology of sinus node disorders

Congenital Heart Disease

- *Pre surgery*
  - Left atrial isomerism
  - Left juxtaposition of atrial appendages
  - Ebstein
- *Post surgery*
  - Ebstein
  - Mustard/Senning
  - Fontan /tCPC
  - Sinus venosus ASD
  - Arterial switch
  - Tetralogy of Fallot
  - Sinus venosus ASD

“Idiopathic”

Genetic

- SCN5A
- HCN4
- RYR2
- CASQ2
- Lamin A/C
- Ankyrin B
- Caveolin 3
- PRKAG2
- LQT
- NKX 2-5

Sinus Node Dysfunction

- Class 1
  - Sinus node dysfunction with correlation of symptoms during age-inappropriate bradycardia (B)
- Class IIa
  - Asymptomatic sinus bradycardia in children and CHD with resting rate < 40 bpm or pauses in ventricular rate > 3 s (C)
  - Sinus node dysfunction with intra-atrial reentrant tachycardia with the need for antiarrhythmic drugs when other therapeutic options, such as catheter ablation, are not possible (C)
  - Congenital heart disease and impaired haemodynamics due to sinus bradycardia or loss of AV synchrony (C)
- Class IIb
  - Asymptomatic sinus bradycardia in the adolescent with CHD with resting rate < 40 bpm or pauses in ventricular rate > 3 s (C)

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Europace
doi:10.1093/europace/eut082
EHRA/AEPC CONSENSUS STATEMENT
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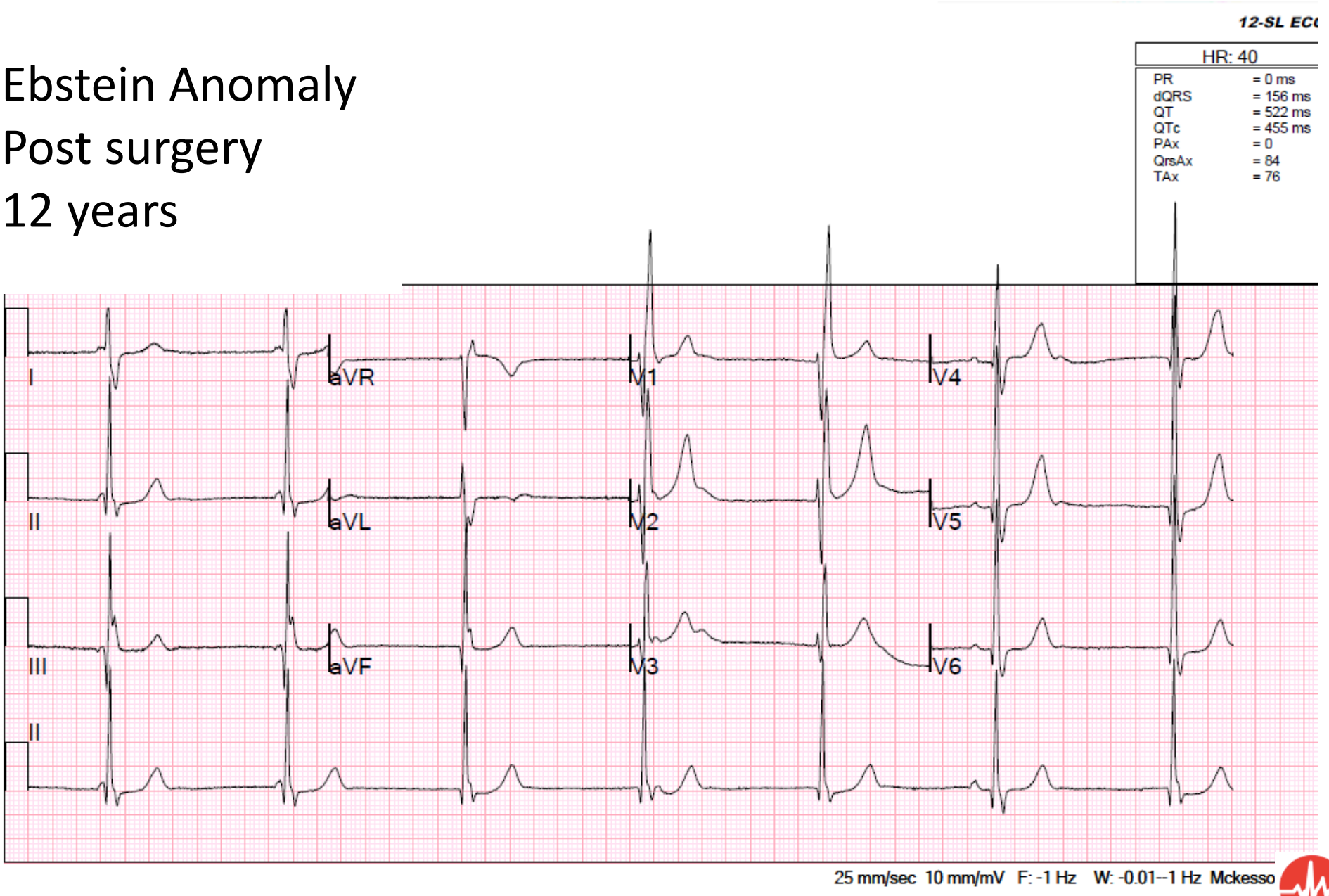
Ebstein Anomaly
Post surgery
12 years



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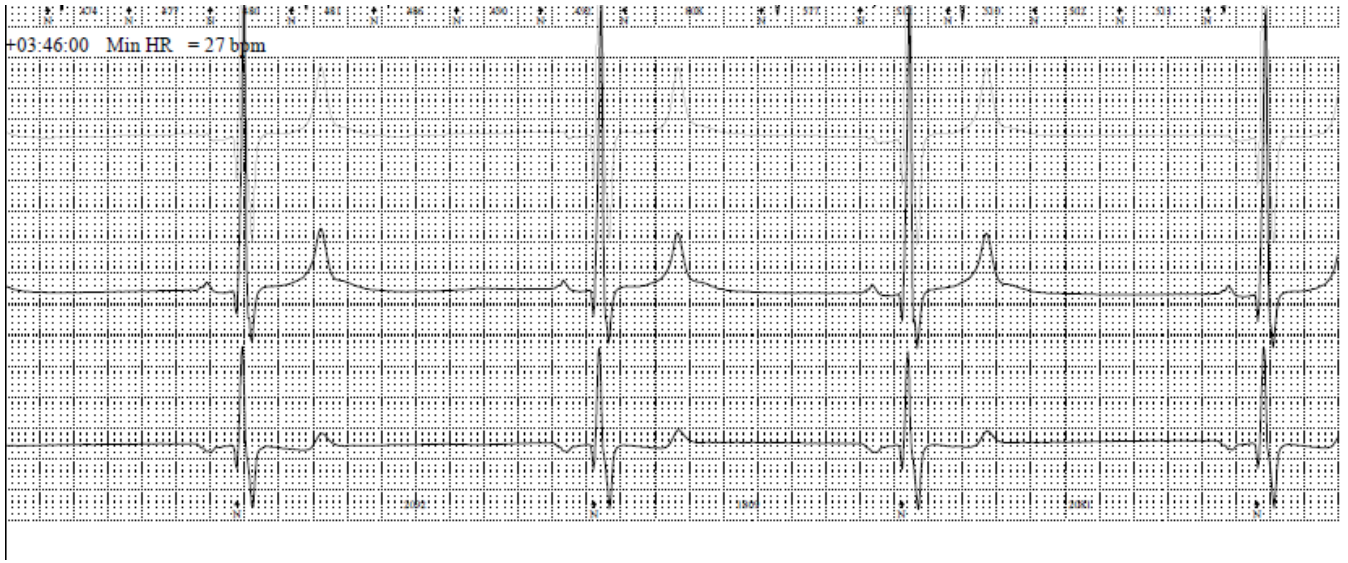

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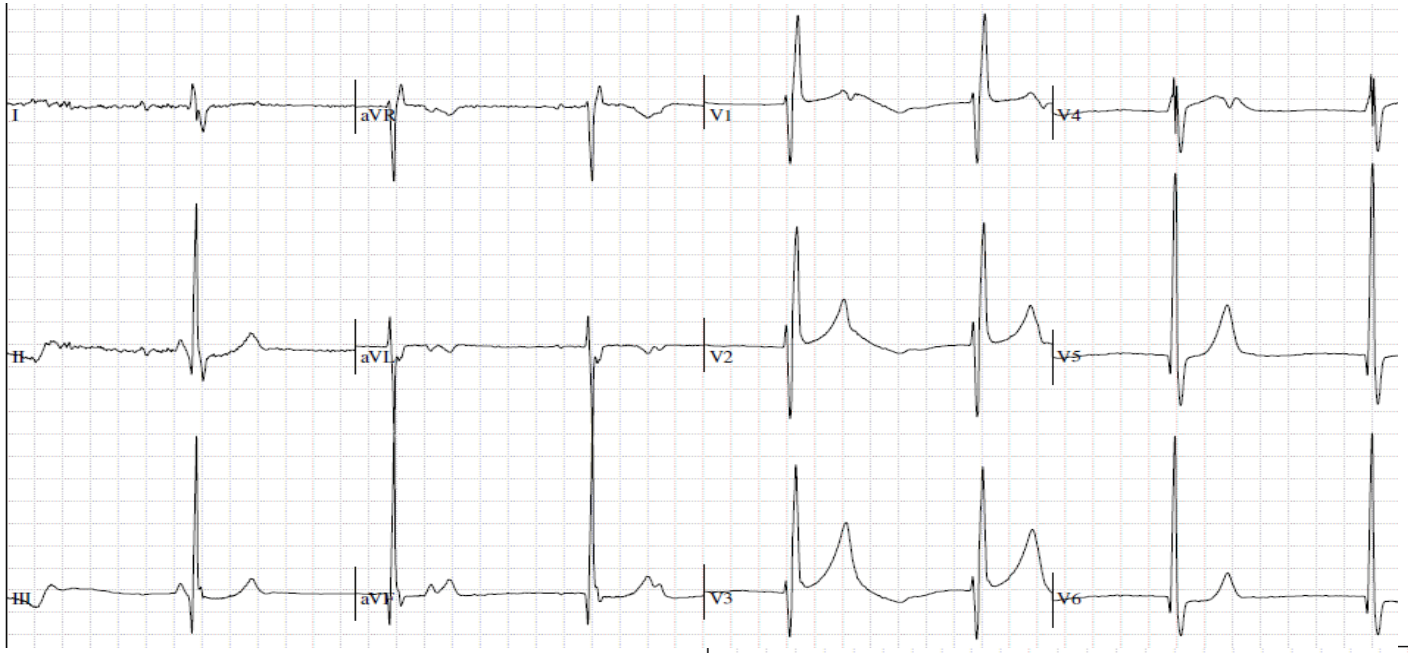

Pre-exercise

EXERCISE
STAGE 2
1:25

PAED ARR
1.7 mph
12.0 %

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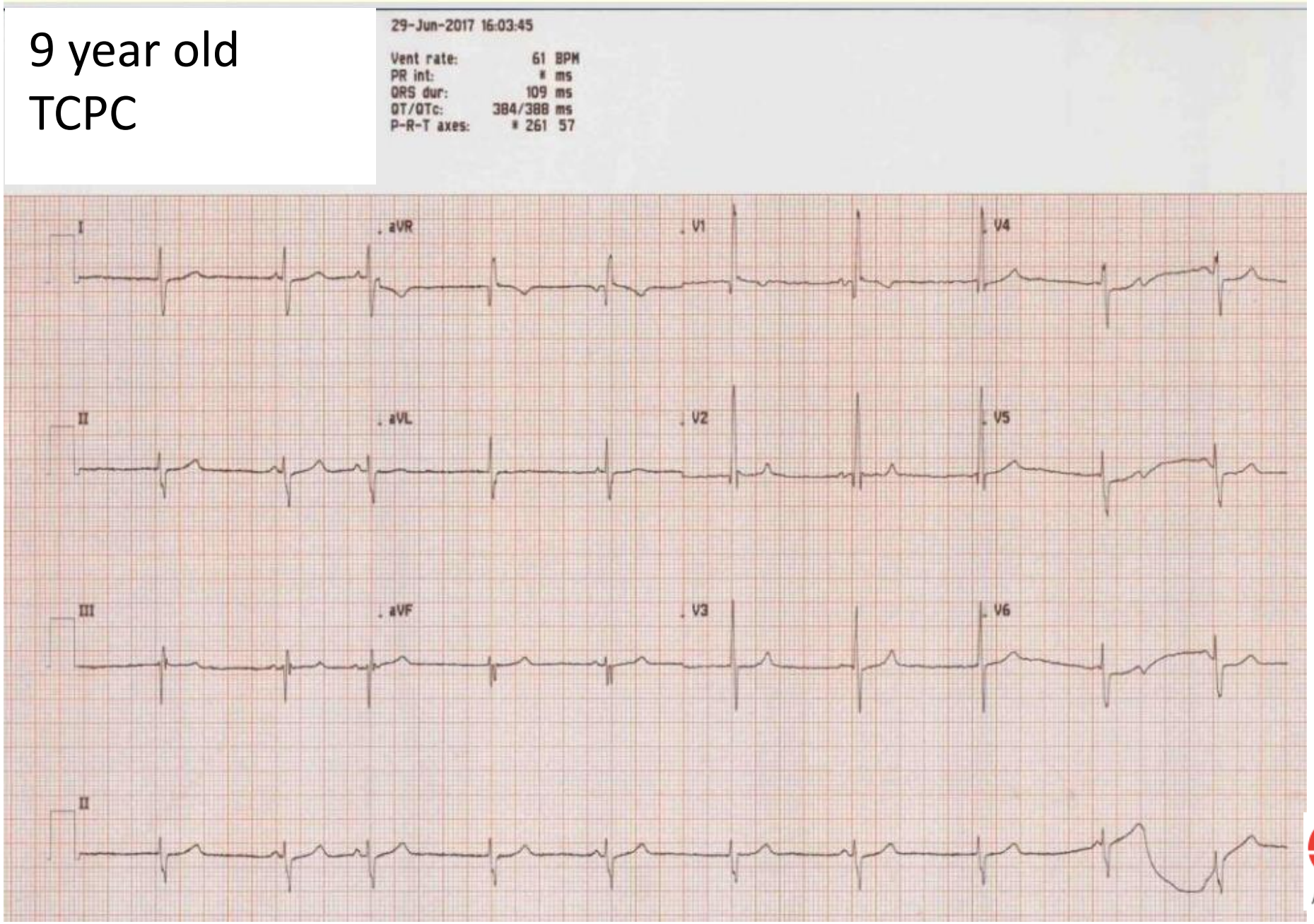
9 year old TCPC



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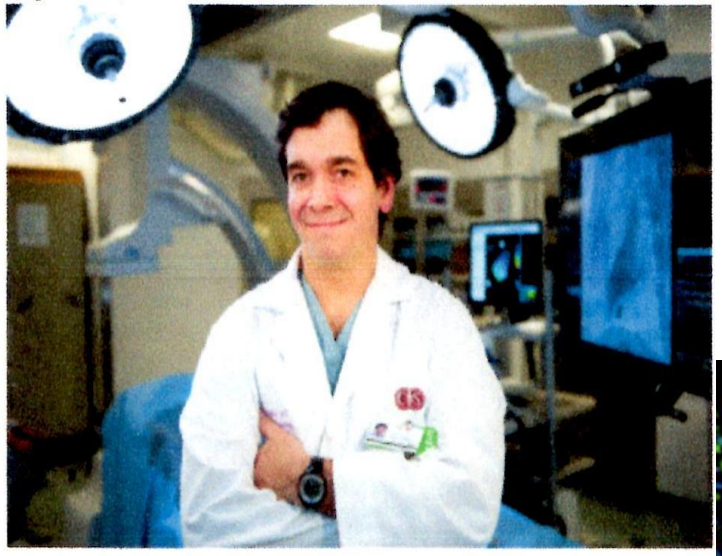
Summary

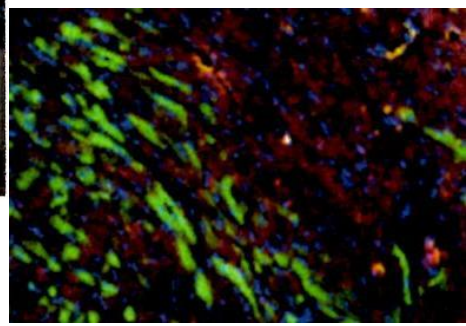
- Current guidelines are just a “guide” based on limited evidence and we need to consider many of our cases individually
- Indications may change as our pacing techniques develop as risk benefit ratio will change
- In CHD indications change with our understanding of consequences of surgical techniques
- Indications may change with understanding of genetic conditions

A biological pacemaker is on its way

Wednesday, 13 Aug 2014 11:54



Eugenio Cingolani



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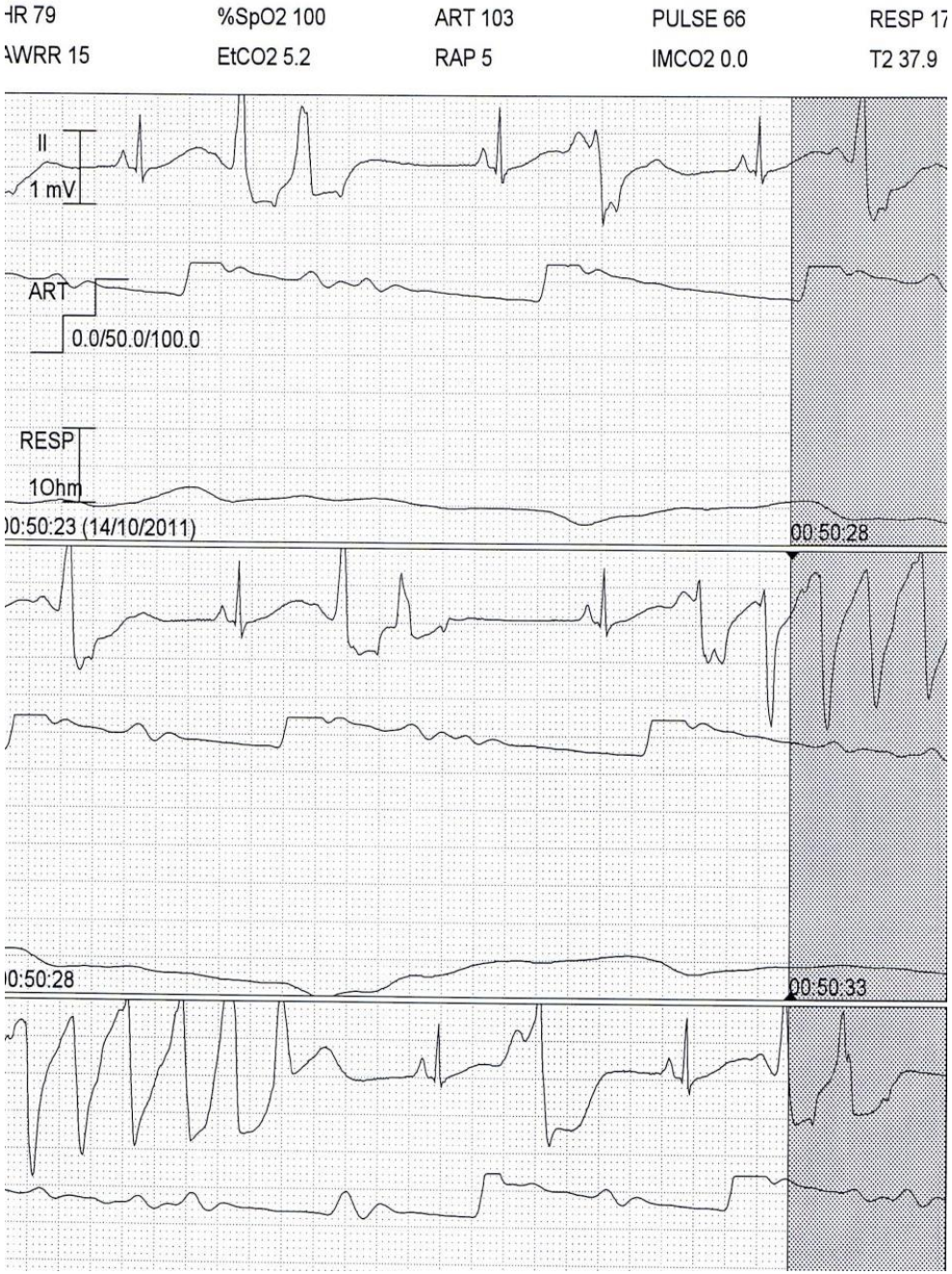
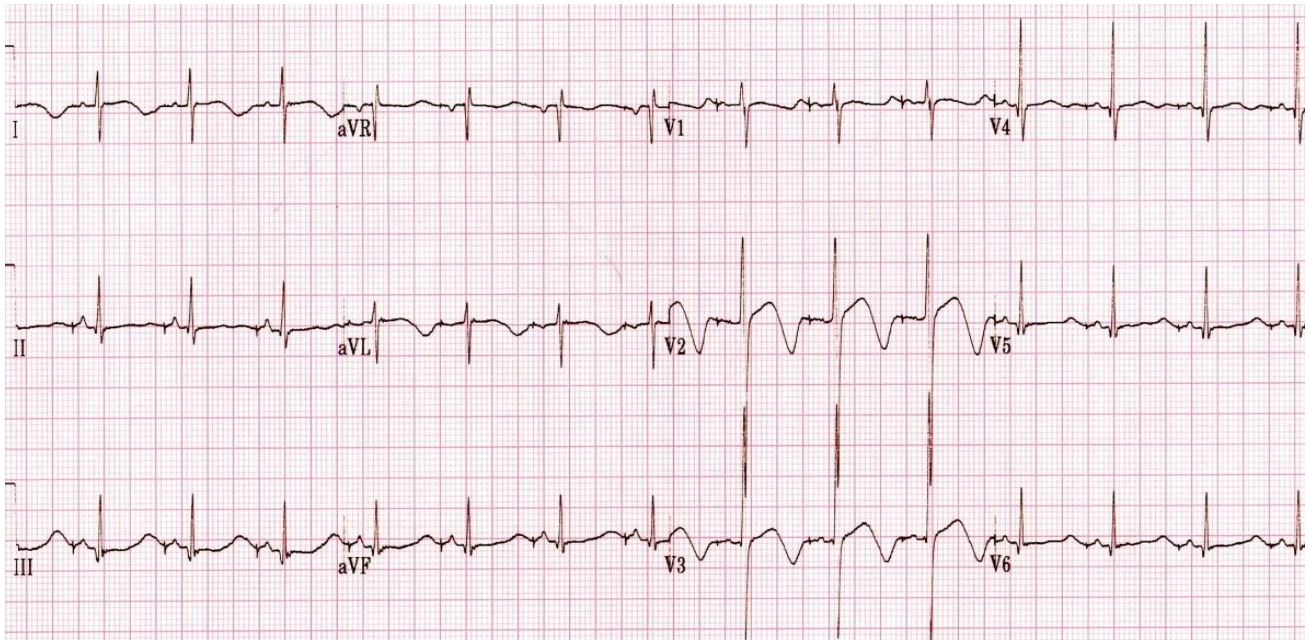
Atrial pacing to stabilise Long QT child with recurrent VT/storm





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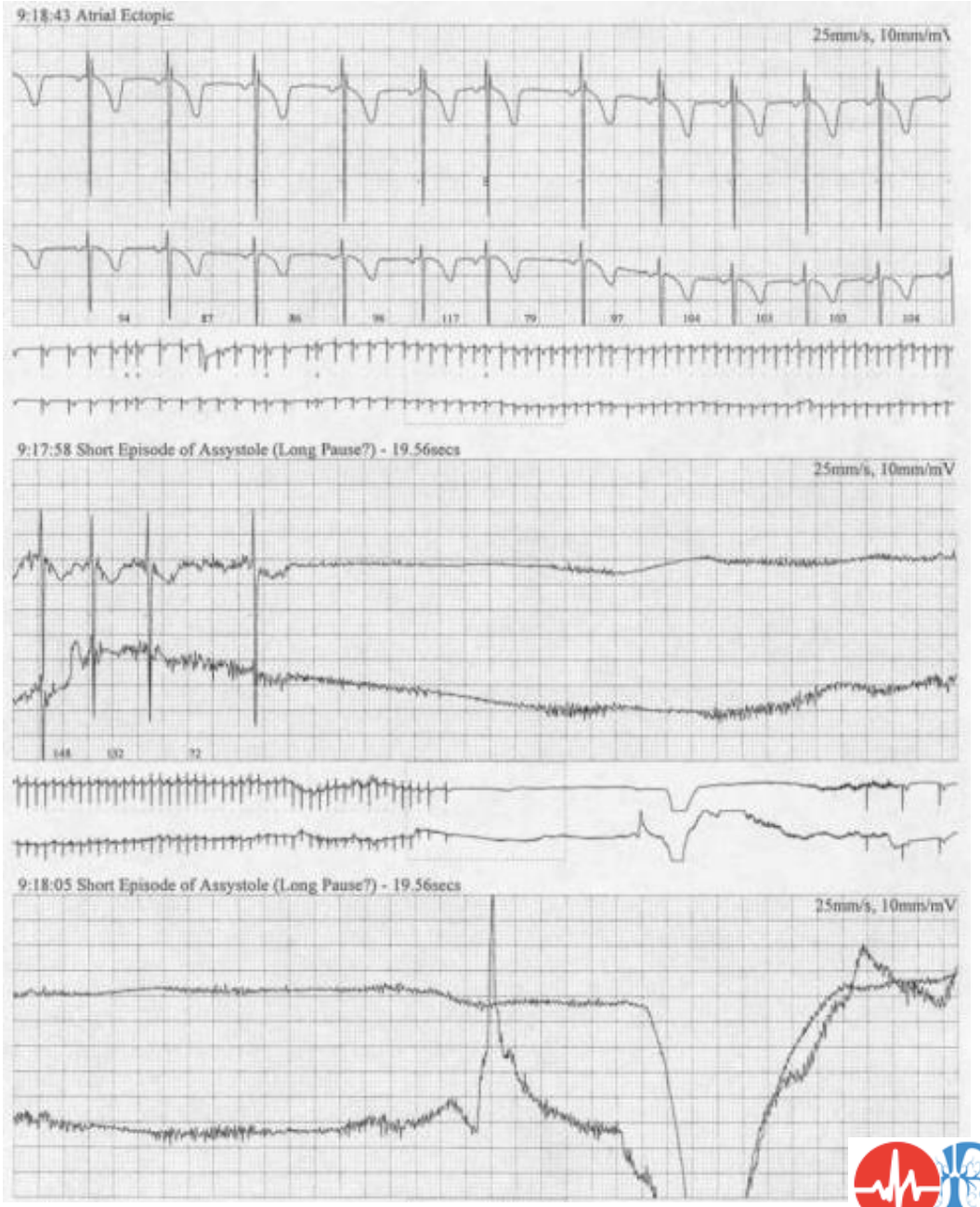
Neurocardiogenic Syncope

- Significantly symptomatic patients in who prolonged asystole can be demonstrated spontaneously or at tilt-table testing (C)

- Class IIb



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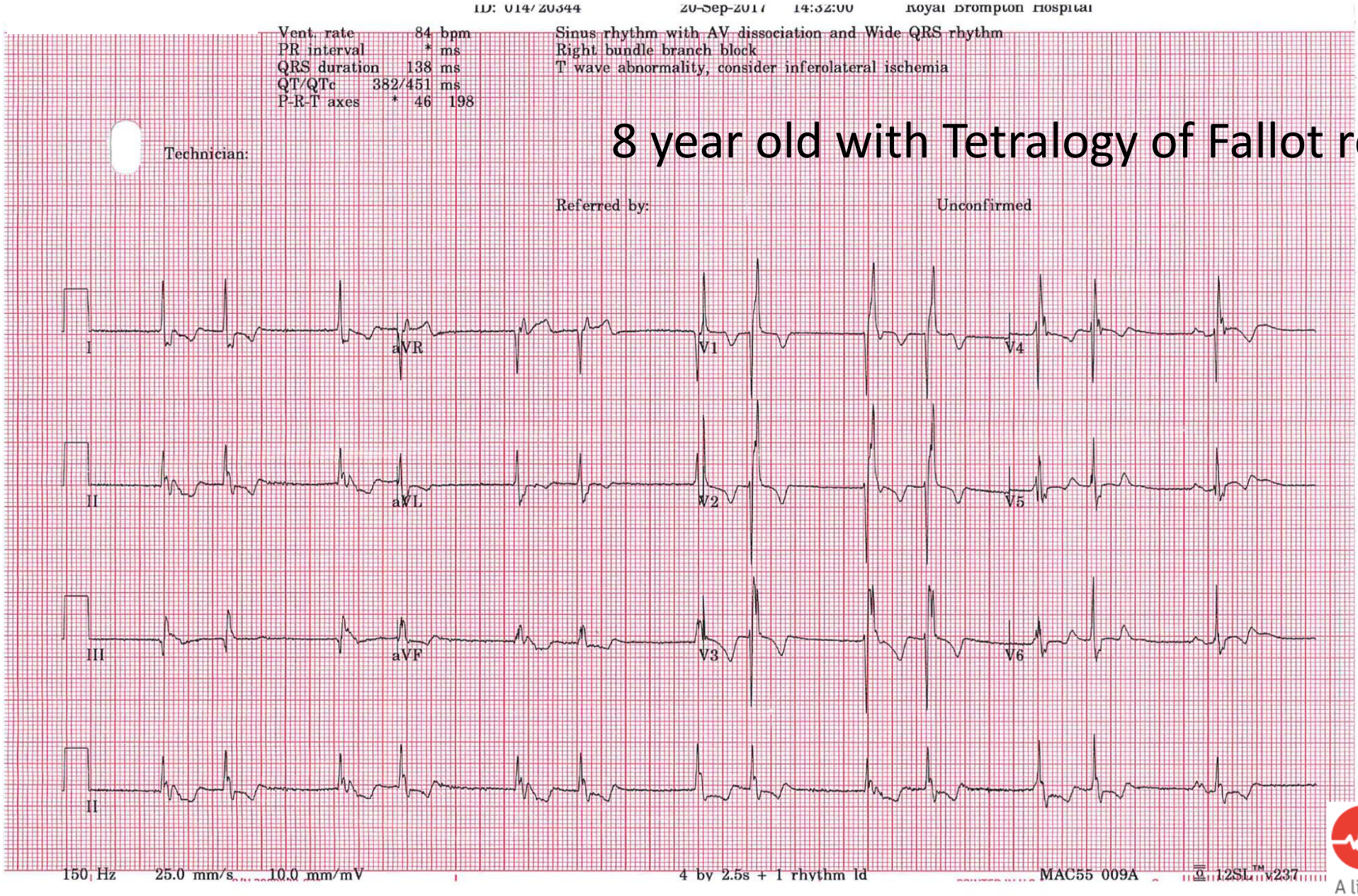
8 year old with Tetralogy of Fallot repair



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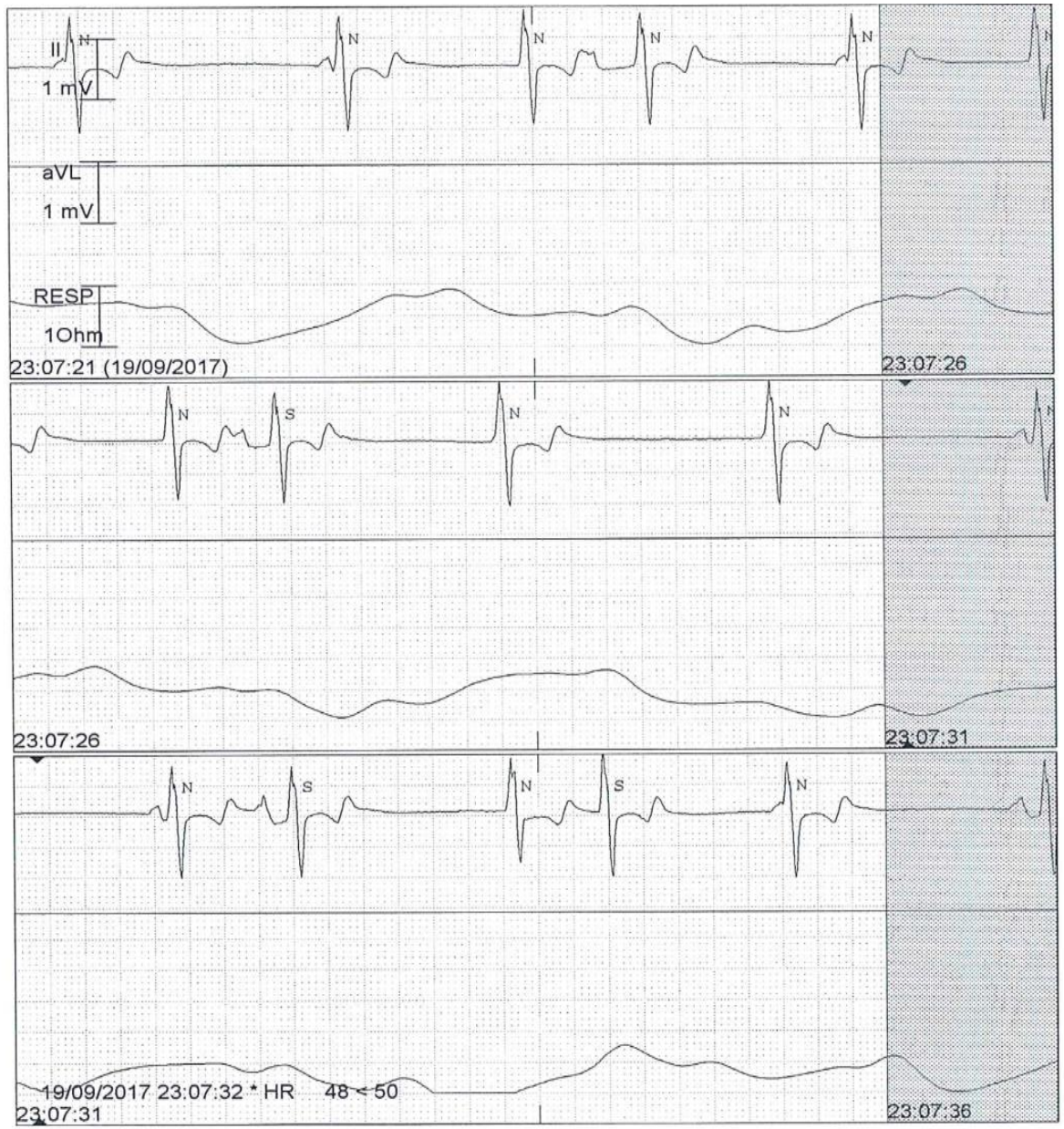

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