Predictors and management of ventricular dysfunction in paced patients

L. Környei
Hungarian Pediatric Heart Center

7th February 2017 11:15 –12:30 PANEL: Complete AV Block from Fetus to Adolescent

Topics

I. Prevalence, mechanism of DCM in CCAVB

II. Influence of the ventricular pacing site on LV function *(predictors)*

III. CRT/Biventricular pacing in children *(management)*

Prevalence of DCM in CCAVB

- DCM/HF developed in 8 of 32 **(25%)** patients who underwent PMI but in none of 6 patients who did not *(Tsujii, Circ J 2016)*
- postnatal DCM was present in 35 cases of 186 **(18.8%)** and among the 35 cases, 9 (26%) had PMI and 14 (40%) died *(Levesque, Autioimmun Rev 2015)*
- 325 children with cardiac NL, 40 **(12%)** cardiac-related deaths, with 37 from cardiomyopathy *(Izmirly. Circulation 2011)*
- impaired LV function in 17 **(12%)** of 141 cases in a multicenter study. *(Eliasson, Circulation 2011)*
- **7-10%** of chronically RV-paced patients develop heart failure and that up to **13%** have depressed LV function combined with LV dilatation. *Moak J Am Coll Cardiol 2011, Gebauer Eur Heart J 2009, Vatasescu Europace 2007*

Etiology of DCM in CCAVB

Table 1. Cardiac Manifestations of Neonatal Lupus[2,3,5]
Congenital complete AV block (CCAVB) (irreversible)
1st degree AV block (may be reversible)
2nd degree AV block (may be reversible)
Dilated cardiomyopathy (from autoimmune injury and/or chronic RV pacing) (can occur in the absence of CCAVB)
Endocardial fibroelastosis (EFE)
Sinus bradycardia or sick sinus syndrome (possibly from autoimmune injury to I_{CaL}, I_{CaT})
QT prolongation
AV valve insufficiency (from chordal rupture)

Horigome, Circulation Journal Vol.80, May 2016

LV dysfunction at F-UP was associated with abnormal LV function before PMI

(Eliasson, Circulation 2011

Etiology of DCM in CCAVB

- pathological studies demonstrated antibodies, complements, and signs of inflammation or fibrosis throughout the myocardium, suggesting the importance of **“autoimmun”** mechanisms

Ambrosi, *Exp Cell Res* 2014

- there is a convincing number of reports demonstrating that chronic right ventricular **(RV) single-site pacing** can cause left ventricular (LV) dysfunction and DCM through ventricular dyssynchrony

Tsujii, *Circ J* 2016
Gebauer, Eur Heart J 2009
van Geldorp Heart Fail Rev 2011

Etiology of DCM in CCAVB

- Chronic **RV pacing, rather than the aetiology of AV block**, has been identified as an independent risk factor for development of LV dilatation and dysfunction

Karpawich, Pacing Clin Electrophysiol 1999

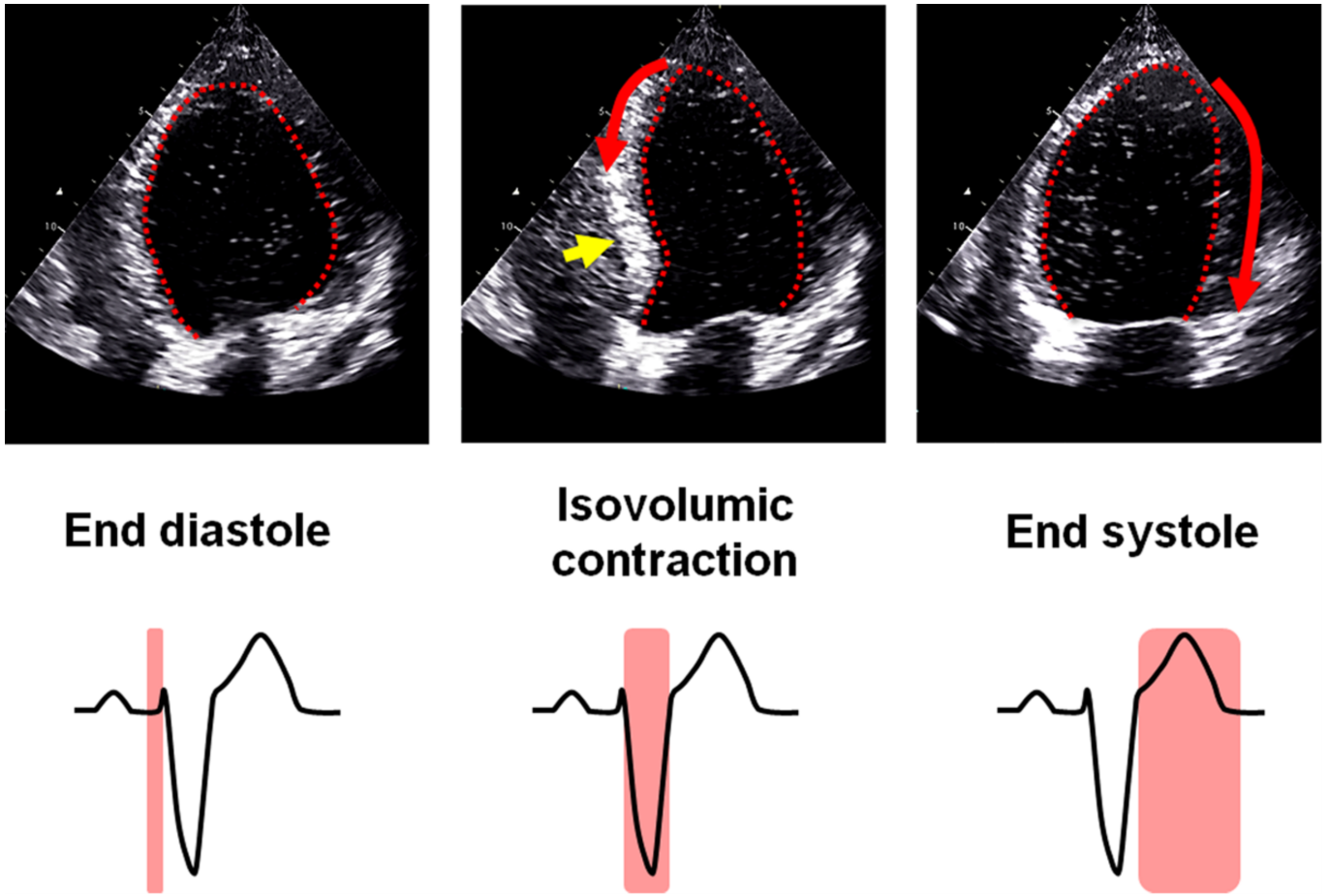
Gebauer, Eur Heart J 2009

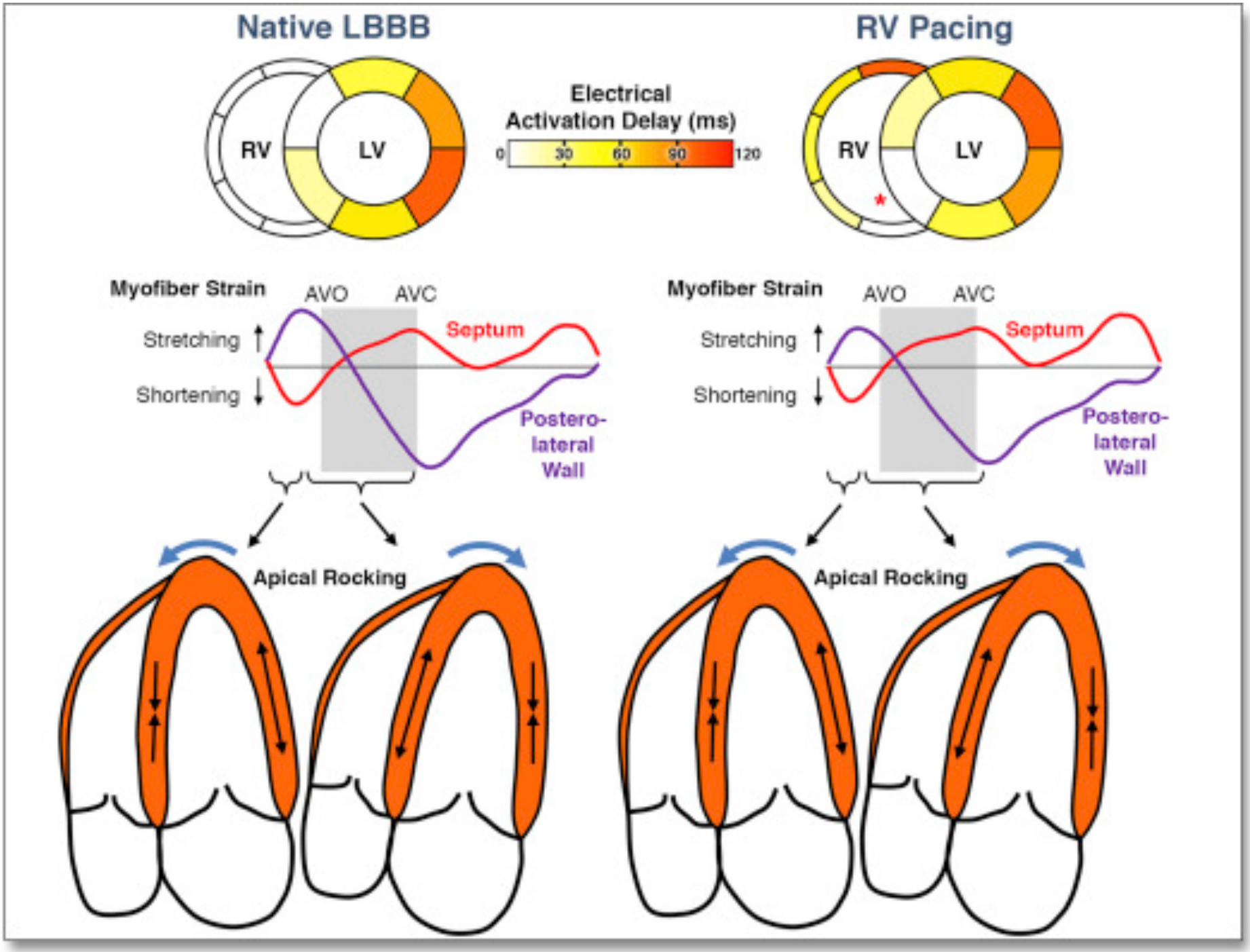
- implication of RV single-site pacing in the development of DCM is supported by the fact that
  - **DCM may develop** after PMI in patients with **originally normal LV** function
  - **improvement or resolution** of LV dysfunction/heart failure **after upgrading** the pacemaker to the CRT system or changing the pacing site to the LV
- **maternal autoantibody status did not significantly influence** LV function or LV dilatation score

van Geldorp Heart Fail Rev 2011

Mechanisms of DCM Development by RV Pacing

Eur Heart J Cardiovasc Imaging. 2015;17(3):262-269. doi:10.1093/ehjci/jev288




Native LBBB
RV
LV
Electrical
Activation Delay (ms)
0
30
60
90
120
RV Pacing
RV
LV
Myofiber Strain
AVO
AVC
Stretching
Shortening
Septum
Postero-
lateral
Wall
Apical Rocking
Myofiber Strain
AVO
AVC
Stretching
Shortening
Septum
Postero-
lateral
Wall
Apical Rocking

Influence of the ventricular pacing site on LV function

Heart

Impact of the permanent ventricular pacing site on left ventricular function in children: a retrospective multicentre survey

Irene E van Geldorp, Tammo Delhaas, Roman A Gebauer, Patrick Frias, Maren Tomaske, Mark K Friedberg, Svjetlana Tisma-Dupanovic, Jan Elders, Andreas Früh, Fulvio Gabbarini, Petr Kubus, Viera Illikova, Sabrina Tsao, Andreas Christian Blank, Anita Hiippala, Thierry Sluysmans, Peter Karpawich, Sally-Ann Clur, Xavier Ganame, Kathryn K Collins, Gisela Dann, Jean-Benoît Thambo, Conceição Trigo, Bert Nagel, John Papagiannis, Annette Rackowitz, Jan Marek, Jan-Hendrik Nürnberg, Ward Y Vanagt, Frits W Prinzen, Jan Janousek and for the Working Group for Cardiac Dysrhythmias and Electrophysiology of the Association for European Paediatric Cardiology

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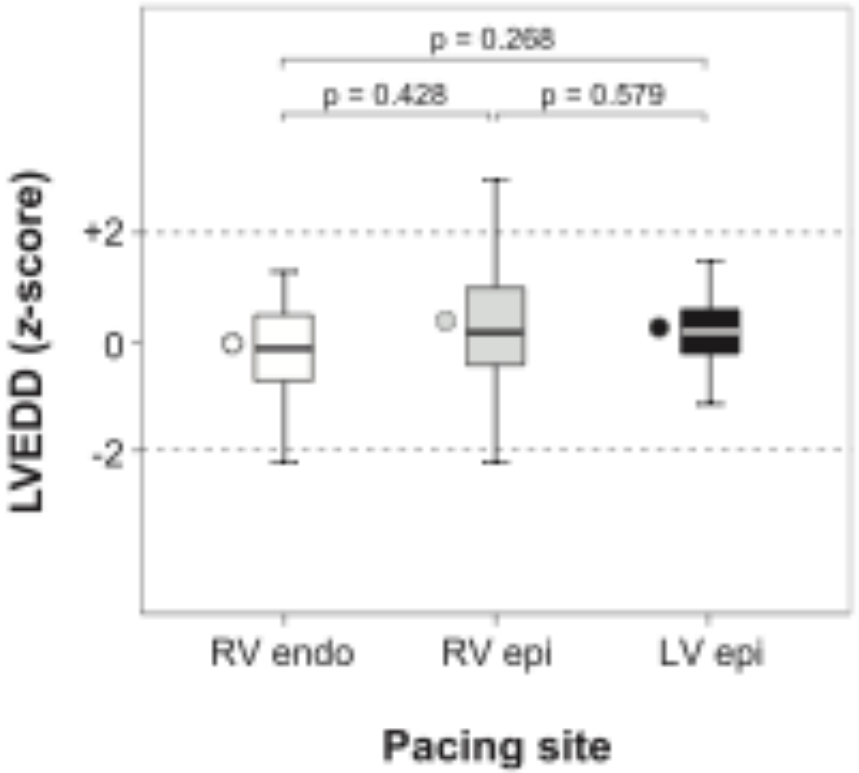
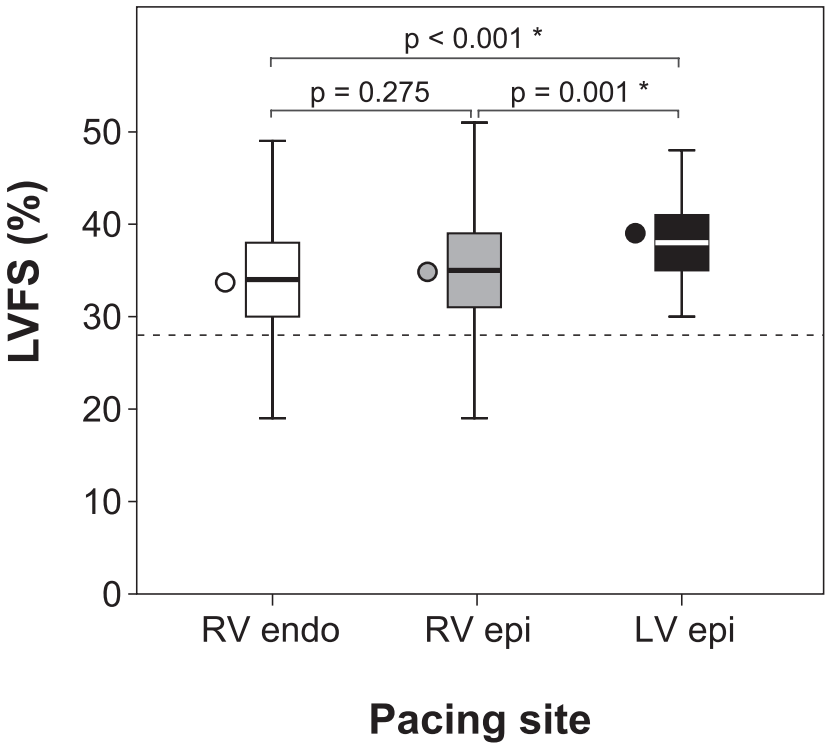
Heart

Impact of the permanent ventricular pacing site on left ventricular function in children: a retrospective multicentre survey





Heart

Impact of the permanent ventricular pacing site on left ventricular function in children: a retrospective multicentre survey

it is predominantly the pacing site that affects LV function, as reflected by

(1) a higher LVFS in the LV- paced group than in the RV-paced groups;

(2) pacing site being the only significant factor influencing LVFS

(3) dissimilar incidence of patients with LVFS<28% between the pacing site group

Circulation Journal
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http://www.j-circ.or.jp

ORIGINAL ARTICLE
Pediatric Cardiology and Adult Congenital Heart Disease

High Incidence of Dilated Cardiomyopathy After Right Ventricular Inlet Pacing in Patients With Congenital Complete Atrioventricular Block

Nobuyuki Tsujii, MD; Aya Miyazaki, MD; Heima Sakaguchi, MD, PhD; Koji Kagisaki, MD; Tetsuya Yamamoto, MD; Michio Matsuoka, MD; Yuriko Shima, MD; Hajime Ichikawa, MD, PhD; Hideo Ohuchi, MD, PhD

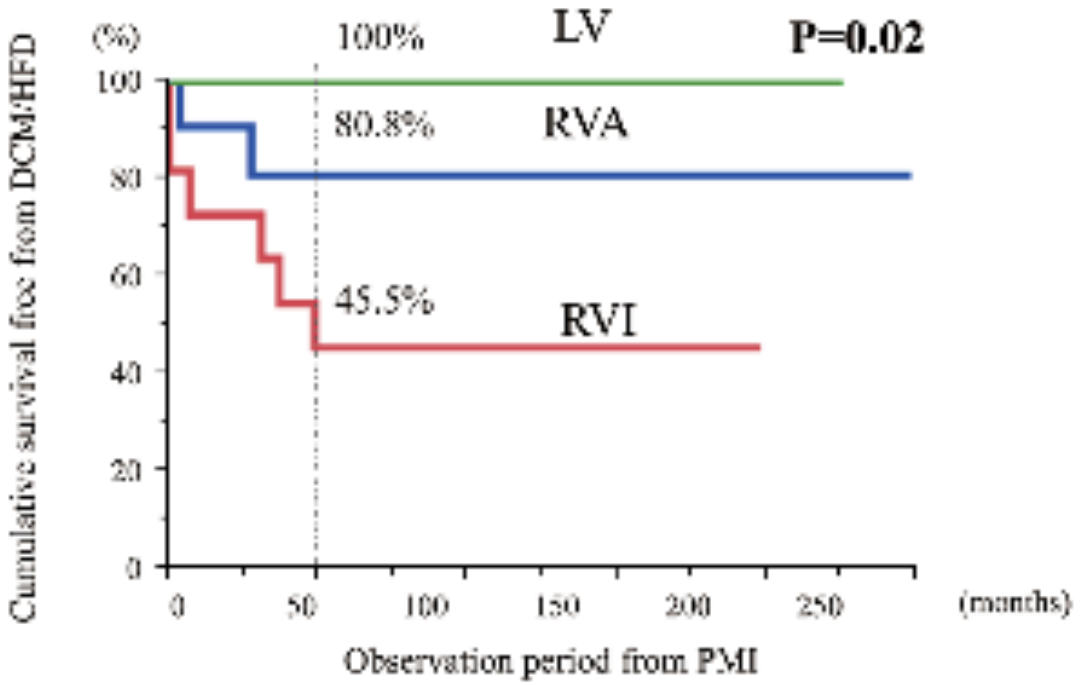

Figure 3. Kaplan-Meier survival curves against the development of DCM/HFD in CCAVB patients after PMI. The cumulative probability of avoidable DCM/HFD at 50 months is 100% with LV pacing, 80.8% with RVA pacing, and 45.5% with RVI pacing (P=0.02). No patient developed DCM or died after 50 months of PMI. CCAVB, congenital complete atrioventricular block; DCM/HFD, dilated cardiomyopathy/heart failure death; PMI, pacemaker implantation; LV, left ventricle; RV, right ventricle; RVA, RV apex; RVI, RV inlet.

Predictors of DCM in CCAVB
preservation of LV function by LV pacing

- During LV (free wall) pacing, the total duration of activation is **prolonged similarly** to that during RV free wall pacing, reflected by a similar QRS duration.

- LV pacing activates the LV lateral wall before the septum and RV lateral wall, **preventing paradoxical** septal movement and resulting in better haemodynamic performance

- above and **beyond synchrony** (reflected by the total duration of activation), the **sequence of activation** is a **major determinant** of cardiac pump function

Management of DCM in CCAVB

Revision of Pacing Site/Mode

Table 2. Examples of CCAVB in Children With DCM Who Underwent Revision of Pacing Site/Mode

Case no.	Author	Year	Original pacing site (age)	Revised PM site/mode (age)	Before PM revision	After PM revision, clinical outcome
1	Moak et al[10]	2006	RV (0.2 years)	CRT (6 years)	LVEF 30%	LVEF 59%
2	Moak et al[10]	2006	RV (3 years)	CRT (15.5 years)	LVEF 20%	LVEF 60%
3	Moak et al[10]	2006	RV (5.5 years)	CRT (17 years)	LVEF 12%	LVEF 66%
4	Vanagt et al[11]	2007	RV free wall (1 day)	LV apex (2 years)	LVSF 20%	LVSF 36%
5	Kurosaki et al[12]	2008	RV (5 days)	LV (5 years)	–	Alive (for 10.5 years), LVSF 38%, CTR 54%, BNP 17 pg/ml
6	Kurosaki et al[12]	2008	RV (10 days)	CRT (4 years)	–	Alive (for 4.3 years), LVSF 20%, CTR 54%, BNP 23 pg/ml
7	Kurosaki et al[12]	2008	RV (1 day)	LV	–	Alive (for 11.2 years), LVSF 31%, CTR 55%, BNP 56 pg/ml
8	Kurosaki et al[12]	2008	RV (5 days)	CRT-P (6 months)	–	Died (7 months) from DCM
9	Gebauer et al[8]	2009	RV free wall (neonatal)	CRT (3.4 years)	NYHA class IV, LVSF 30%	Deterioration, LVSF 10%
10	Matsuhisa et al[13]	2014	RV lateral (0.2 years)	LV apex (3.8 years)	LVSF 24%, BNP 18.2 pg/ml	LVSF 41%, BNP 10.2 pg/ml
11	Matsuhisa et al[13]	2014	LV base (1.2 years)	LV apex (4.7 years)	LVSF 14%, BNP 327 pg/ml	LVSF 48%, BNP 21.1 pg/ml
12	Horigome et al[15]	2014	RV (neonatal)	CRT-P (16 months)	CTR 70%, BNP 6,520 pg/ml	CTR 55%, BNP 28 pg/ml
13	Ellesøe et al[16]	2014	RV outflow (31 days)	CRT-P (16 months)	LVSF 10%	LVSF 33%
14	Tsujii et al[4]	2016	RV inflow	LV apex	–	Alive (for 7 years), NYHA class I
15	Tsujii et al[4]	2016	RV apex	CRT	–	Alive (for 8 years), NYHA class I

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CLINICAL RESEARCH
Cardiac electrophysiology

Left ventricular pacing in neonates and infants with isolated congenital complete or advanced atrioventricular block: short- and medium-term outcome

Massimo Stefano Silvetti[1]*, Duccio Di Carlo[2], Antonio Ammirati[1], Silvia Placidi[1], Corrado Di Mambro[1], Lucilla Ravà[3], and Fabrizio Drago[1]

[1]Arrhythmia Unit and Syncope Unit, Department of Paediatric Cardiology, Bambino Gesù Children's Hospital, IRCCS, Via Torre di Palidoro 1, 00050 Palidoro-Rome, Italy; [2]Cardiac Surgery Unit, Bambino Gesù Children's Hospital, IRCCS, 00100 Rome, Italy; and [3]Epidemiology Unit, Bambino Gesù Children's Hospital, IRCCS, 00100 Rome, Italy

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CLINICAL RESEARCH
Cardiac electrophysiology

Left ventri … d infants with isolat … dvanced atrioventr … lium-term outcome

Massimo Stefano … , Silvia Placidi[1],
Corrado Di Mam …

[1]Arrhythmia Unit and Syncope Unit, … o 1, 00050 Palidoro-Rome, Italy;
[2]Cardiac Surgery Unit, Bambino Ges … ospital, IRCCS, 00100 Rome, Italy

Received 9 September 2013; accepted

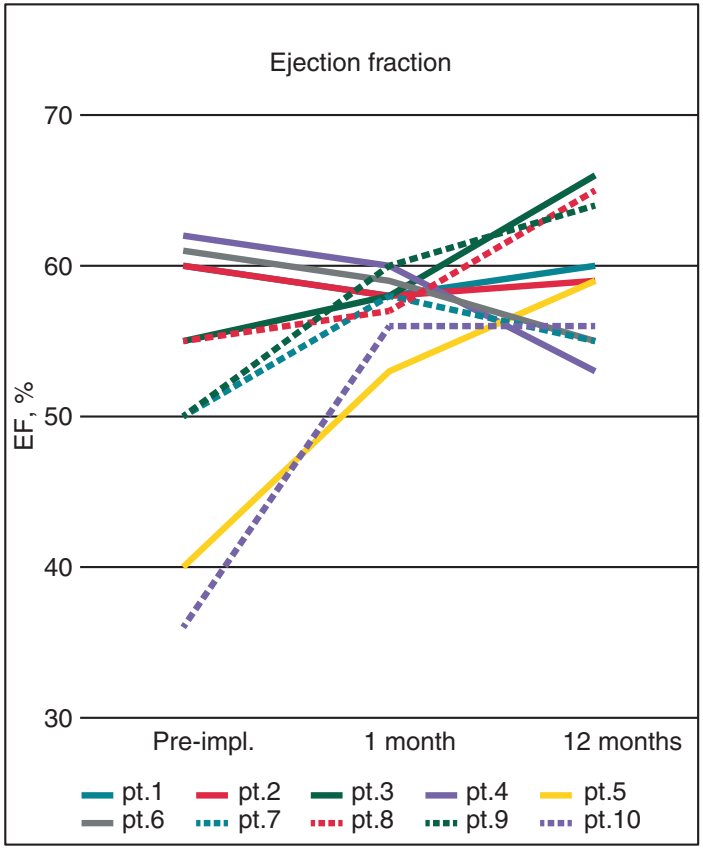

Figure 2 Changes in LV EF. pt., patient; pre-impl., before pacemaker implantation. Median (range): pre-impl. 55% (36–62%); 1 month 58% (53–60%); 12 months 59% (53–66%). Kendall's coefficient 0.35, $P = 0.4$. See text for further details.

Summary I.

- Development of DCM in CCAVB children is frequently associated with pre-PMI LV dysfunction possibly caused by an autoimmune mechanism

- However, it can occur after chronic RV single-site pacing, even in patients with completely normal echocardiography before PMI

Summary II.

- tailored follow-up of patients at risk and
- accurate monitoring of LV function before and after PMI

MANDATORY

- for detection of deterioration of LV dysfunction at an early stage (+revision of pacing site)

Thank you !