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EUROPEAN CLOSED CONSENSUS CONFERENCE ON

DIAGNOSIS OF ARRHYTHMOGENIC CARDIOMYOPATHY



Florence, Italy · 9-10 June 2022 · NH Collection Hotel Palazzo Gaddi

INTRO SPEECH

Criteria for diagnosis of arrhythmogenic cardiomyopathy (ACM) were first proposed in 1994 and revised in 2010 by an International Task Force. Although the International Task Force criteria demonstrated a good accuracy for diagnosis of the original right ventricular phenotype (arrhythmogenic right ventricular cardiomyopathy), they lacked sensitivity for identification of the expanding phenotypic spectrum of ACM, which includes left-sided variants and did not incorporate late-gadolinium enhancement findings by cardiac magnetic resonance. The lack of specific diagnostic criteria for the left ventricular phenotype of ACM has resulted in clinical under-recognition of patients with phenotypes other than the original ARVC over the 4 decades since the disease discovery. The development of the 2020 upgrading of diagnostic criteria (“the Padua criteria”) by International experts was based on the evolving clinical experience with the expanding spectrum of ACM phenotypes and needed to fill the diagnostic gap of the previous 1994 and 2010 TF guidelines. The key upgrade was the incorporation of tissue characterization findings by cardiac magnetic resonance for noninvasive detection of late-gadolinium enhancement/myocardial fibrosis that are determinants for characterization of arrhythmogenic biventricular and left ventricular phenotypes. and to provide a codification for future translational and clinical research. The 2020 Padua criteria are heavily dependent on cardiac magnetic resonance, which has become mandatory to characterize the ACM phenotype and to exclude other diagnoses. New criteria regarding left ventricular depolarization and repolarization ECG abnormalities and ventricular arrhythmias of left ventricular origin were also provided. The 2020 Padua criteria aimed to improve the diagnosis of ACM by providing new criteria for diagnosis of biventricular and ALVC phenotypes, particularly by means of the incorporation of CMR tissue characterization findings as well as LV depolarization and repolarization ECG abnormalities and ventricular arrhythmias. This new diagnostic approach needs to be validated by clinical studies on large patient populations. Preliminary data confirm that the clinical use of the Padua criteria substantially impacts the diagnostic accuracy and permits a comprehensive identification of the phenotypic variety of ACM, mostly by virtue of demonstration of RV and LV LGE/myocardial fibrosis by CMR. The application of the upgraded diagnostic criteria in real-world clinical practice will be crucial for future refinement and correlation with therapeutic outcomes.

The aim of this Conference is to refine the “Padua criteria” and to reach a large international consensus for the proposal of the 2022 European Task Force Criteria (“refined Padua criteria”), as a result of an interactive discussion between European panellists chosen for their expertise in the basic science or clinical manifestations of the disease.

The conference will run over two full days and will consist of a series of short presentations followed by interactive round table discussion focused on key aspects of the disease diagnosis, differential diagnosis with phenocopies and characterization of etiologic variants of the disease.

President of the Meeting
Prof. Domenico Corrado

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European Reference Network
GUARD-Heart

PRESIDENT OF THE CONFERENCE

Prof. Domenico Corrado

Inherited Arrhythmogenic Cardiomyopathies and Sports Cardiology Unit,
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Cristina Basso (IT)
Chiara Bucciarelli Ducci (UK)
Domenico Corrado (IT)
William McKenna (UK)
James Moon (UK)
Jeanette Schulz-Menger (DE)
Antonis Pantazis (UK)
Gaetano Thiene (IT)
Thomas Wichter (DE)
Arthur Wilde (NL)

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PROGRAMME

THURSDAY, 9 JUNE 2022 – MORNING

08.30 a.m.
09.00 a.m.

D. Corrado (Italy)
Introduction and Conference outline
Fondazione Internazionale Menarini - Welcome Addresses

SESSION I – EVOLVING CRITERIA FOR DIAGNOSIS OF ACM

Chairpersons:

G. Thiene (Italy)
W. J. McKenna (UK)
D. Corrado (Italy)

09.00 a.m.
09.20 a.m.

G. Thiene (Italy)
Story telling of ARVC

09.20 a.m.
09.40 a.m.

W. J. McKenna (UK)
The 1994 and 2010 Diagnostic Criteria for ARVC

09.40 a.m.
10.00 a.m.

D. Corrado (Italy)
The “Padua Criteria“ and a “3-step“ approach to diagnosis of ACM

10.00 a.m.
10.30 a.m.

Panel Discussion

10.30 a.m.
11.00 a.m.

Coffee Break

THURSDAY, 9 JUNE 2022 – MORNING

SESSION II – RIGHT VENTRICULAR PHENOTYPE (I): ECG ABNORMALITIES, VENTRICULAR ARRHYTHMIAS AND DIFFERENTIAL DIAGNOSIS

Chairpersons:	J. Tfelt (Denmark) A. Anastasakis (Greece) B. Bauce (Italy)
11.00 a.m. 11.15 a.m.	S.Sharma (UK) Basal ECG
11.15 a.m. 11.30 a.m.	B. Bauce (Italy) Holter monitoring, exercise testing and SAECG
11.30 a.m. 11.45 a.m.	E. Gandjbakhch (France) Ventricular arrhythmias and electrophysiologic study
11.45 a.m. 12.00 p.m.	C. Blomström Lundqvist (Sweden) Differential diagnosis with RVOT tachycardia
12.00 p.m. 12.15 p.m.	A. Pelliccia (Italy) Differential diagnosis with the athlete's heart
12.15 p.m. 01.00 p.m.	Panel Discussion
01.00 p.m. 02.00 p.m.	<i>Lunch</i>

THURSDAY, 9 JUNE 2022 – AFTERNOON

SESSION III – RIGHT VENTRICULAR PHENOTYPE (II): MORPHO-FUNCTIONAL AND STRUCTURAL CRITERIA

Chairpersons:	C. Blomström Lundqvist (Sweden) T. Wichter (Germany) A. Pantazis (UK)
02.00 p.m. 02.15 p.m.	A. Pantazis (UK) Echocardiographic criteria
02.15 p.m. 02.30 p.m.	T. Wichter (Germany) Angiographic criteria
02.30 p.m. 02.45 p.m.	C. Bucciarelli-Ducci (UK) CMR morpho-functional criteria
02.45 p.m. 03.00 p.m.	A. Cipriani (Italy) CMR structural criteria
03.00 p.m. 03.15 p.m.	C. Basso (Italy) EMB criteria
03.15 p.m. 03.30 p.m.	M. Perazzolo Marra (Italy) CMR structural vs EMB criteria
03.30 p.m. 04.00 p.m.	Panel Discussion
04.00 p.m. 04.30 p.m.	<i>Coffee break</i>

THURSDAY, 9 JUNE 2022 – AFTERNOON

SESSION IV – LEFT VENTRICULAR PHENOTYPE (I): MORPHO-FUNCTIONAL, STRUCTURAL CRITERIA AND DIFFERENTIAL DIAGNOSIS

Chairpersons:	J. Schulz-Menger (Germany) J. Moon (UK) M. Perazzolo Marra (Italy)
04.30 p.m. 04.45 p.m.	M. Perazzolo Marra (Italy) CMR features (I)
04.45 p.m. 05.15 p.m.	J. Jiménez Jaimez (Spain) CMR features (II)
05.15 p.m. 05.15 p.m.	J. Moon (UK) CMR features (III)
05.15 p.m. 05.30 p.m.	D. Corrado (Italy) Similarities and differences with “Arrhythmogenic Dilated Cardiomyopathy”
05.30 p.m. 05.45 p.m.	J. Schulz-Menger (Germany) Similarities and differences with post-myocarditis scar
05.45 p.m. 06.00 p.m.	C. Bucciarelli-Ducci (UK) Similarities and differences with cardiac sarcoidosis
06.00 p.m. 06.30 p.m.	Panel Discussion
06.30 p.m.	<i>End of the day</i>

PROGRAMME

FRIDAY, 10 JUNE 2022 – MORNING

SESSION V – LEFT VENTRICULAR PHENOTYPE (II): ECG ABNORMALITIES AND VENTRICULAR ARRHYTHMIAS, DIFFERENTIAL DIAGNOSIS WITH NEUROMUSCULAR CARDIOMYOPATHIES AND OTHER

Chairpersons:	M. Kharlap (Russia) P. Seferovic (Serbia) C. De Asmundis (Belgium)
08.30 a.m. 08.45 a.m.	A. Zorzi (Italy) ECG and ventricular arrhythmias
08.45 a.m. 09.00 a.m.	J. Tfelt (Denmark) RBBB morphology ventricular tachycardia
09.00 a.m. 09.15 a.m.	J. Schulz-Menger (Germany) Similarities and differences with muscular dystrophy-related cardiomyopathy
09.15 a.m. 09.30 a.m.	J. Moon (UK) Similarities and differences with Lamin A/C-related neuromuscular cardiomyopathy
09.30 a.m. 09.45 a.m.	A. Cipriani (Italy) Similarities and differences with Filamin C-related neuromuscular cardiomyopathy
09.45 a.m. 10.00 a.m.	T. Wichter (Germany) Similarities and differences with LV congenital aneurysms
10.00 a.m. 10.30 a.m.	Panel Discussion
10.30 a.m. 11.00 a.m.	<i>Coffee break</i>

FRIDAY, 10 JUNE 2022 – MORNING

SESSION VI – GENETICS AND GENOTYPE-PHENOTYPE CORRELATIONS

Chairpersons:	R. Jurcut (Romania) L. Monserrat (Spain) A. Wilde (The Netherlands)
11.00 a.m. 11.15 a.m.	L. Monserrat (Spain) Desmosomal genes
11.15 a.m. 11.30 a.m.	K. Pillichou (Italy) Non-desmosomal genes
11.30 a.m. 11.45 a.m.	R. Jurcut (Romania) PKP-related ACM
11.45 a.m. 12.00 p.m.	B. Bauce (Italy) DSP-related ACM
12.00 p.m. 12.15 p.m.	A. Wilde (The Netherlands) PLN-related ACM
12.15 p.m. 12.30 p.m.	J. Jiménez Jaimez (Spain) DES-related ACM
12.30 p.m. 01.00 p.m.	Panel Discussion
01.00 p.m. 02.00 p.m.	<i>Lunch</i>

FRIDAY, 10 JUNE 2022 – AFTERNOON

SESSION VII –2022 EUROPEAN TASK FORCE CRITERIA (REFINED “PADUA CRITERIA”) FOR DIAGNOSIS OF ACM

Chairpersons:	C. Bucciarelli-Ducci (UK) C. Basso (Italy) D. Corrado (Italy)
02.00 p.m. 02.15 p.m.	C. Basso (Italy) Current Pathologic Criteria for Diagnosis of ACM: common histopathologic denominator between genetic and acquired disease variants
02.15 p.m. 02.30 p.m.	D. Corrado (Italy) The revised ACM phenotypes: implications for classification of cardiomyopathies
02.30 p.m. 03.00 p.m.	Panel Discussion Clinical and imaging criteria for RV phenotype
03.00 p.m. 03.30 p.m.	Panel Discussion Clinical and imaging criteria for LV phenotype
03.30 p.m. 04.00 p.m.	Panel Discussion Diagnosis of phenotypic variant (ARVC, BIV or ALVC)
04.00 p.m. 04.30 p.m.	Panel Discussion Diagnostic role of molecular genetics
04.30 p.m. 05.00 p.m.	Panel Discussion Differential diagnosis: phenocopies or etiologic disease variants? Towards an etiologic classification of ACM including both genetic and acquired disease
05.00 p.m.	<i>Concluding remarks</i>

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GENERAL INFORMATION

Event reserved to Faculty Members.

MEETING VENUE

The venue for the Meeting will be:

Hotel NH Collection Firenze Palazzo Gaddi – Via Del Giglio 9 – 50123 Florence (Italy) + 39 055 2398095

SECRETARIAT DURING THE MEETING

The Secretariat will be open at the following times:

Thursday, 9 June, from 08.00 a.m. to 06.30 p.m.

Friday, 10 June, from 08.00 a.m. to 05.00 p.m.

OFFICIAL LANGUAGE

The official language of the Meeting will be English.

TECHNICAL FACILITIES

Facilities will be available for computer presentations and overhead projections.

A business center with PC (Powerpoint for Windows) will be available for check and preview of presentations. It is essential that speakers take their presentation to the business center at least one hour before the session starts.

The center will be open at the following times:

Thursday, 9 June, from 08.00 a.m. to 06.30 p.m.

Friday, 10 June, from 08.00 a.m. to 05.00 p.m.

LUNCHES AND COFFEE BREAKS

Lunches and coffee breaks will be served in the Meeting area.

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