

Cardiomyopathy Registry

Case Report Form

2 Patient Characteristics

FAMILY No: ____

PATIENT (family member) No: ____

(e.g. Family 1, Patient 1; Family 1, Patient 2)

Patient: **Index case* (Proband)** **Relative** Unknown

(e.g. *first or most typically affected member of the family)

Relative: **Affected** **Not affected** (not necessarily to be entered)

2.1 Basic Information

Type of patient Inpatient Outpatient

Familial disease* No Yes
 Not possible to determine
 Not evaluated

(e.g. *At least one other relative in family with definite diagnosis of cardiomyopathy.)

2.2 Enrolment

Date of birth (mm/yyyy) |_|_| |_|_|_|_|_|

Date of first evaluation at the centre |_|_| |_|_| |_|_|_|_|_|

2.3 Physical Characteristics

Gender Male Female

Height _____ cm

Weight _____ kg

Body mass index _____ (Automatically calculated)

Body surface area _____ (Automatically calculated)

2.4 Diagnosis

Date of diagnosis of cardiomyopathy |_|_| |_|_|_|_|_| (mm/yyyy)

Cardiomyopathy type

- Hypertrophic
- Dilated
- Arrhythmogenic
- Restrictive
- Non-compact
- Tako-tsubo

Syndromic disease

- No
- Yes

If yes, please specify:

Reason for diagnosis

- Incidental
- Symptoms
- Sudden death/cardiac arrest
- Family screening
- Other
- Not recorded

2.5 Comorbidities / Risk Factors

Family history of Sudden Death

(in 1 or more relatives under 40 years old)

- No
- Yes
- Unknown

Hypertension

- No
- Yes

If yes, date of diagnosis

|_|_| |_|_| |_|_|_|_|

Diabetes mellitus

- No
- Type I
- Type II

If Type II, insulin-dependent

- No
- Yes

Hyperlipidaemia/dyslipidaemia

- No
- Yes

Physical activity

- None (no exercise or exercise for < 3 hours/week for <2 years)
- Occasionally (< 3 hours/week for ≥ 2 years)
- Regularly (> 3 hours/week for ≥ 2 years)
- Intensely (> 7 hours/week for ≥ 2 years)
- Unknown

Alcohol use, in units None < 1/day 1/day
 2-3/day >4/day Unknown

1 unit = 1 glass of wine or beer or any other alcoholic beverage with an equivalent amount of alcohol

Smoking No Current
 Former (> 1 month) Unknown

Smoking: Current = patient regularly smokes a tobacco product(s) one or more times per day or has smoked in the 30 days prior to this admission. Former = patient has stopped smoking tobacco products more than 30 days before this admission. Indicate if the patient has a history confirming any form of tobacco use in the past. This includes cigarettes, cigar and/or pipe.

Drug use No Cocaine Others

If others, please specify _____

Renal impairment No Yes

Depressiveness No Yes

Skeletal muscle impairment No Yes

Chronic obstructive pulmonary disease No Yes

Anaemia No Yes

Comments on environmental factors, other diseases: _____

2.6 Symptoms

Presence of symptoms No Yes

Age at first symptom Known Unknown

If known |__|__| years

Syncope events
 None
 Vagal or non-cardiac syncope
 Suspected arrhythmic/cardiogenic syncope
 Mechanism uncertain
 Not evaluated

Number of suspected arrhythmic/cardiogenic syncope _____

Age at last arrhythmic/cardiogenic syncope |__|__| years

Anginal chest pain No Yes

NYHA functional class NYHA I NYHA II NYHA III
 NYHA IV Not available/possible

Extreme lethargy No Yes

Palpitations No Yes

Ankle oedema No Yes

Orthopnea No Yes

Paroxysmal nocturnal dyspnea No Yes

History of Atrial Fibrillation No Yes

If yes, type of AF Paroxysmal Persistent
 Permanent Unknown

History of sustained ventricular tachycardia No Yes

History of resuscitated ventricular fibrillation/cardiogenic arrest No Yes

History of stroke No TIA
 Stroke Not recorded

History of AV block No Yes

History of BBB No Yes

3 Investigations

3.1 Electrocardiogram

ECG

Not Performed Performed

|_|_| |_|_| |_|_|_|_|

Date of ECG

Rhythm

- Sinus rhythm
- Atrial fibrillation Atrial flutter
- Atrial paced AV paced
- SR V paced AF V paced
- Ventricular tachycardia
- Other Not evaluated

Frequent premature beats

No Yes

Heart rate

_____ bpm

QT interval

_____ msec

PR interval

_____ ms

QRS duration

_____ ms

QRS axis

_____ degrees

Atrioventricular block

- No 1st degree
- 2nd degree 3rd degree

Bundle branch block

- No Incomplete RBBB
- RBBB Incomplete LBBB
- LBBB Other

Negative T waves

No Yes

ST depression

No Yes

ST elevation

No Yes

Maximum R in praecordial

_____ mm

Maximum S in praecordial

_____ mm

Maximum R in limbs

_____ mm

Maximum S in limbs

_____ mm

Preexcitation No Yes

Abn Q-waves No Yes

3.2 Echocardiogram

Echocardiogram Not performed Performed

|_|_|_| |_|_|_| |_|_|_|_|_|_|

Date of echocardiogram

LVEDD _____ mm

Estimated LVEDD¹ (Henry formula) _____ mm (*automatically calculated*)

% of the estimated LV end-diastolic diameter² (Henry formula) _____ (*automatically calculated*)

LVESD _____ mm

LV ejection fraction (Simpson's biplane) _____ %

Fractional shortening _____ %

LVEDV (LV End Diastolic Volume) _____ ml

LVESV (LV end systolic volume) _____ ml

Maximum LV thickness _____ mm

LV septal thickness diastole _____ mm

LV posterior wall thickness diastole _____ mm

Left atrium diameter _____ mm

Left atrial area _____ cm²

Pericardial effusion No Yes

Endocavitary thrombi No Yes

Pattern of LV hypertrophy

No hypertrophy Asymmetrical septal

Concentric Apical

Other/not classified Not evaluated

¹ LVEDD calculated as follows: $((45,3 * ([BSA]^{1/3})) - (0,03 * [Age]) - 7,2)$

² Calculated as follows: $([LVEDd]/[LVEDdEstHenry]) * 100$

- RV dilation No Yes
- No Yes
- RV global systolic dysfunction
- RV hypertrophy No Yes
- Maximum RV wall thickness _____ mm
- New wall motion abnormality No Yes

3.3 Doppler Diastolic Function Assessment

- Doppler** Not performed Performed
- Date of Doppler echo** |_|_| |_|_| |_|_|_|_|
- Mitral E-wave** _____ m per second
- E-wave deceleration time** _____ m per second
- Mitral A-wave** _____ m per second
- TDI lateral mitral annulus peak E velocity m/s (E or Ea)** _____ m per second
- E/Ea (automatic calculation required to generate E/Ea (ie. Mitral E-wave m/s divided by Ea m/s))**
- Diastolic dysfunction (grade)** Normal
 Grade I (impaired relaxation)
 Grade II (pseudo normal)
 Restrictive
 Not evaluated
- Left ventricular outflow tract gradient (resting)** _____ mmHg
- _____ mmHg
- Maximum provoked (by any technique) peak left ventricular outflow tract gradient**
- Aortic regurgitation None Mild
 Moderate Severe
- Mitral regurgitation** None Mild
 Moderate Severe
- Tricuspid regurgitation** None Mild
 Moderate Severe
- Systolic Pulmonary Artery pressure** _____ mmHg
 Not possible/evaluated

- Aortic stenosis None Mild
 Moderate Severe
- Mitral stenosis None Mild
 Moderate Severe
- Valve replacement No Yes
 If yes, replaced valves Mitral Aortic Tricuspid
 Pulmonary

3.4 Cardiac Magnetic Resonance Imaging MRI scan

- Not performed Normal
 Abnormal Inconclusive

Date of the MRI

|_|_| |_|_| |_|_|_|_|

Late gadolinium enhancement (LGE)

- No Yes
 Not possible/not evaluated

LGE location, specify

LV end-diastolic volume _____ ml

LV end-systolic volume _____ ml

LV ejection fraction _____ % (*absolute number*)

Maximum LV thickness by CMR _____ mm

Left ventricular mass (LVM) _____ g

Pattern of hypertrophy

- No hypertrophy Septal
 Concentric Eccentric
 Apical Other
 Unknown

3.5 Holter

Holter Not performed Performed

Date of the Holter |_|_| | |_|_| | |_|_|_|_|_|

Non-sustained ventricular tachycardia No Yes Unknown

Rhythm

- Sinus rhythm throughout
- Paroxysmal atrial fibrillation in sinus rhythm
- Atrial fibrillation throughout
- Sinus rhythm & V pacing
- Permanent atrial fibrillation & V pacing
- AV pacing
- Conduction defect (AVB 1-2-3; sinus dysfunction)

3.6 Exercise Test

Exercise test Not performed Performed

If yes, date (dd/mm/yyyy) |_|_| | |_|_| | |_|_|_|_|_|

Exercise type Bicycle Treadmill

Medication on day of exercise test

- No
- Beta-blocker
- Verapamil or Diltiazem
- Other
- Unknown

If Other, please specify _____

Systolic blood pressure, baseline _____ mmHg

Systolic blood pressure, max achieved _____ mmHg

Systolic blood pressure, peak exercise _____ mmHg

Absolute workload achieved _____ Watts

_____ METS

Peak heart rate _____ bpm

Ventricular arrhythmia No Yes

Supraventricular arrhythmia No Yes

3.7 Laboratory

3.7.1 At first evaluation at the time of entry in the registry (last value available)

Collection date (dd/mm/yyyy)	_ _ _ _ _ _ _ _ _ _ _ _ _ _ _ _ _
Haemoglobin	_____ <input type="checkbox"/> mmol/L <input type="checkbox"/> g/dl
White blood cells	_____ <input type="checkbox"/> $10^3/\text{mm}^3$ <input type="checkbox"/> $10^9/\text{L}$
Platelets	_____ <input type="checkbox"/> $10^3/\text{mm}^3$ <input type="checkbox"/> $10^9/\text{L}$
Creatine kinase	_____ <input type="checkbox"/> IU/L
Troponin I	_____ ng/ml
Troponin T	_____ ng/ml
Creatinine	_____ <input type="checkbox"/> $\mu\text{mol}/\text{L}$ <input type="checkbox"/> mg/dL
Total cholesterol	_____ <input type="checkbox"/> mmol/L <input type="checkbox"/> mg/dL
Fasting glucose	_____ <input type="checkbox"/> mmol/L <input type="checkbox"/> mg/dL
Sodium	_____ <input type="checkbox"/> mEq/L <input type="checkbox"/> mmol/L
Potassium	_____ <input type="checkbox"/> mEq/L <input type="checkbox"/> mmol/L
Uric acid	_____ <input type="checkbox"/> $\mu\text{mol}/\text{L}$ <input type="checkbox"/> mg/dL
HbA1c	_____ <input type="checkbox"/> % <input type="checkbox"/> mmol/mol
Nitrogen measured by:	
BUN (<i>Blood Urea Nitrogen</i>)	_____ <input type="checkbox"/> mmol/L <input type="checkbox"/> mg/dL
Urea	_____ <input type="checkbox"/> mmol/L <input type="checkbox"/> mg/dL
BNP	_____ <input type="checkbox"/> pmol/L <input type="checkbox"/> pg/mL
NT-proBNP	_____ <input type="checkbox"/> pmol/L <input type="checkbox"/> pg/mL
T4	_____ <input type="checkbox"/> pmol/L <input type="checkbox"/> ng/dL
TSH	_____ <input type="checkbox"/> mIU/L

3.8 Signal averaged ECG

Date of signal averaged ECG:

Normal (0/3) 1/3 criteria

2/3 criteria

3/3 criteria

Not performed

1. *Filtered QRS duration (fQRS) ≥ 114 ms*
2. *Duration of terminal QRS $< 40 \mu V$ (low-amplitude signal duration) ≥ 38 ms*
3. *Root-mean-square voltage of terminal 40 ms $\leq 20 \mu V$*

4 Medication Section

Is the patient on any medication? No Yes

Alpha-blockers	<input type="checkbox"/> No <input type="checkbox"/> Yes
Beta-blockers	<input type="checkbox"/> No <input type="checkbox"/> Sotalol <input type="checkbox"/> Carvedilol <input type="checkbox"/> Bisoprolol <input type="checkbox"/> Metoprolol <input type="checkbox"/> Nebivolol <input type="checkbox"/> Other If other, specify: _____ Dose: _____
Diuretics, oral <i>(Multiple possible answers)</i>	<input type="checkbox"/> No <input type="checkbox"/> Bendrofluazide <input type="checkbox"/> Hydrochlorothiazide <input type="checkbox"/> Indapamide <input type="checkbox"/> Torasemide <input type="checkbox"/> Chlorthalidone <input type="checkbox"/> Furosemide <input type="checkbox"/> Bumetanide <input type="checkbox"/> Other If other, specify: _____ Dose: _____
ACE inhibitors	<input type="checkbox"/> No <input type="checkbox"/> Perindopril <input type="checkbox"/> Ramipril <input type="checkbox"/> Lisinopril <input type="checkbox"/> Enalapril <input type="checkbox"/> Captopril <input type="checkbox"/> Fosinopril <input type="checkbox"/> Other If other, specify: _____ Dose: _____
Angiotensin II receptor blockers	<input type="checkbox"/> No <input type="checkbox"/> Candesartan <input type="checkbox"/> Losartan <input type="checkbox"/> Valsartan <input type="checkbox"/> Other If other, specify: _____ Dose: _____
Mineralocorticoid antagonists (MRAs)	<input type="checkbox"/> No <input type="checkbox"/> Spironolactose <input type="checkbox"/> Eplerenone <input type="checkbox"/> Canrenone <input type="checkbox"/> Other If other, specify: _____ Dose: _____

Antiplatelets <i>(Multiple possible answers)</i>	<input type="checkbox"/> No <input type="checkbox"/> Clopidogrel <input type="checkbox"/> Prasugrel <input type="checkbox"/> Ticagrelor <input type="checkbox"/> Aspirin <input type="checkbox"/> Other
	If other, specify: _____ Dose: _____
Oral anticoagulants	<input type="checkbox"/> No <input type="checkbox"/> Vit K Antagonists <input type="checkbox"/> Dabigatran <input type="checkbox"/> Rivaroxaban <input type="checkbox"/> Apixaban <input type="checkbox"/> Other If other, specify: _____ Dose: _____
Anti-arrhythmic drugs <i>(Multiple possible answers)</i>	<input type="checkbox"/> No <input type="checkbox"/> Amiodarone <input type="checkbox"/> Digitoxin/digoxin <input type="checkbox"/> Flecainide <input type="checkbox"/> Propafenone <input type="checkbox"/> Disopyramide <input type="checkbox"/> Other If other, specify: _____ Dose: _____
Calcium antagonists	<input type="checkbox"/> No <input type="checkbox"/> Diltiazem <input type="checkbox"/> Verapamil <input type="checkbox"/> Other: _____ Dose: _____
Nitrates	<input type="checkbox"/> No <input type="checkbox"/> Yes
Statins	<input type="checkbox"/> No <input type="checkbox"/> Yes
Ivabradine	<input type="checkbox"/> No <input type="checkbox"/> Yes
Direct renin inhibitors	<input type="checkbox"/> No <input type="checkbox"/> Yes
Other drugs, specify: _____	

Comments on medication: _____

5 Other Procedures

5.1 Device Therapy

Cardioverter defibrillator implanted

No Yes

If yes, date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

Reason for cardioverter defibrillator

Primary prophylaxis
 Secondary prophylaxis

Pacemaker implanted

No Yes

If yes, date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

Reason for pacemaker
(Multiple answers possible)

Bradyarrhythmia
 Treatment of left ventricular outflow tract obstruction
 Cardiac resynchronisation therapy
 Other

Comments on device therapy: _____

5.2 Other Procedures

Septal myectomy

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

Alcohol septal ablation

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

Heart transplant

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

Ventricular assist device

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

EP study performed

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

Cardiac ablation

No

Yes

If yes

For ventricular tachycardia

No

Yes

Date (dd/mm/yyyy)

|_|_|_| |_|_|_| |_|_|_|_|_|_|

For atrial fibrillation

No

Yes

Date (dd/mm/yyyy)

|_|_|_| |_|_|_| |_|_|_|_|_|_|

For nodal tachycardia

No

Yes

Date (dd/mm/yyyy)

|_|_|_| |_|_|_| |_|_|_|_|_|_|

Comments on other procedures: _____

7 Arrhythmogenic Cardiomyopathy

7.1 Morphological Criteria

7.1.1 Major echocardiographic Criteria

Regional RV akinesia, dyskinesia or aneurysm and PLAX RVOT $\geq 32 \text{ mm}^3$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Regional RV akinesia, dyskinesia or aneurysm and PSAX RVOT $\geq 36 \text{ mm}^4$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Regional RV akinesia, dyskinesia or aneurysm and RV area fractional change $\leq 33\%$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown

Comments on echocardiographic features: _____

7.1.2 Major Cardiac Magnetic Resonance (CMR) Criteria

Regional RV akinesia, dyskinesia or dyssynchronous RV contraction and ratio of RV end-diastolic volume to BSA in male $>110 \text{ mL/m}^2$, in female $\geq 100 \text{ mL/m}^2$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and RVEF $\leq 40\%$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Regional RV akinesia, dyskinesia or aneurysm and RV area fractional change $\leq 33\%$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown

Comments on CMR features: _____

7.1.3 Major Angiography Criteria

³ Adjusted (BSA) $\geq 19 \text{ mm}^2$

⁴ Adjusted (BSA) $\geq 21 \text{ mm}^2$

Regional RV akinesia, dyskinesia or aneurysm	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
--	--

Comments on RV and LV angio features: _____

7.1.4 Minor Echocardiographic Criteria

Regional RV akinesia or dyskinesia and PLAX RVOT ≥ 29 to < 32 mm ¹⁶	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Regional RV akinesia or dyskinesia and PSAX RVOT ⁵ ≥ 32 to < 36 mm	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Regional RV akinesia or dyskinesia and RV area fractional area change $> 33\%$ to $\leq 40\%$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown

7.1.5 Minor Cardiac Magnetic Resonance (CMR) Criteria

Regional RV akinesia, dyskinesia or dyssynchronous RV contraction and Ratio of RV end-diastolic volume to BSA ≥ 100 to < 110 mL/m ² (male) or ≥ 90 to < 100 mL/m ² (female)	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Regional RV akinesia, dyskinesia or dyssynchronous RV contraction and RV ejection fraction $> 40\%$ to $\leq 45\%$	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown

7.2 Histological Criteria

7.2.1 Major tissue criteria

Fibrofatty replacement of myocardium defined as: residual myocytes $< 60\%$ by morphometric analysis (or $< 50\%$ if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
---	--

7.2.2 Minor tissue criteria

⁵ Corrected for body size [PSAX/BSA] ≥ 18 to < 21 mm/m²

Residual myocytes 60% to 75% by morphometric analysis (or 50% to 65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

- No Yes
 Not evaluated/unknown

Comments on histology: _____

¹⁶ djusted (BSA) ≥ 16 to < 19 mm/m₂

7.3 ECG Criteria

7.3.1 Major ECG criteria

<p>Inverted T waves in right praecordial leads (V1, V2, and V3) or beyond in individuals > 14 years of age (in the absence of complete right bundle branch block QRS ≥ 120 ms)</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown</p>
<p>Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right praecordial leads (V1 to V3)</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown</p>

7.3.2 Minor ECG criteria

<p>Inverted T waves in leads V1 and V2 in individuals older than 14 years of age (in the absence of complete right bundle branch block) or in V4, V5, or V6</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown</p>
<p>Inverted T waves in leads V1, V2, V3, and V4 in individuals older than 14 years of age in the presence of complete right bundle branch block</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown</p>
<p>Late potentials by SAECG in at least one of the three following parameters⁶:</p> <ol style="list-style-type: none"> 3. Filtered QRS duration (fQRS) ≥ 114 ms 4. Duration of terminal QRS < 40 μV (low-amplitude signal duration) ≥ 38 ms 5. Root-mean-square voltage of terminal 40 ms ≤ 20 μV 	<p><input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown</p>

⁶ in the absence of a QRS duration of > 110 ms on the standard ECG

Terminal activation duration of QRS \geq 55 ms measured from the nadir of the S wave to the end of the QRS, including R', in V1, V2, or V3, in the absence of complete right bundle branch block	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
--	--

Comments on ECG or SAECG: _____

7.4 Arrhythmia Criteria

7.4.1 Major Arrhythmia Criteria

Non-sustained or sustained ventricular tachycardia of left bundle branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and a VF and positive in lead a VL)	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
---	--

7.4.2 Minor Arrhythmia Criteria

Non-sustained or sustained ventricular tachycardia of RV outflow configuration, left bundle-branch block morphology with inferior axis (positive QRS in leads II, III, and a VF and negative in lead a VL) or of unknown axis	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Ventricular ectopics (>500/24h if proband)	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown

Comments on arrhythmia: _____

7.5 Family History

7.5.1 Major Family History Criteria

ARVC/D confirmed in a first-degree relative who meets task force criteria	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
---	--

ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Identification of a pathogenic mutation categorized as associated or probably associated with ARVC/D in the patient	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown

7.5.2 Minor Family History Criteria

History of ARVC/D in a first-degree relative in whom it is not possible or practical to determine whether the family member meets task force criteria	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
Premature sudden death (<35 years of age) due to suspected ARVC/D in a first-degree relative.	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown
ARVC/D confirmed pathologically or by task force criteria in second-degree relative	<input type="checkbox"/> No <input type="checkbox"/> Yes <input type="checkbox"/> Not evaluated/unknown

Comments on family history or genetics: _____

8 Genetics Information

Genetic counselling Performed

If yes, who performed the genetic counselling?

No Yes Unknown

- Geneticist
 Genetic Counsellor
 Cardiologist
 Nurse
 Unknown

Genetic testing performed?

No Yes Unknown

Gene(s) tested (use as many lines as genes screened):

GENE	Result(s)		DNA variant	protein variant
_____	benign possibly benign variant of unknown significance (VUS) possibly pathogenic pathogenic	<input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>	_____ _____ _____ _____	_____ _____ _____ _____
_____	benign possibly benign variant of unknown significance (VUS) possibly pathogenic pathogenic	<input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>	_____ _____ _____ _____	_____ _____ _____ _____
_____	benign possibly benign variant of unknown significance (VUS) possibly pathogenic pathogenic	<input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>	_____ _____ _____ _____	_____ _____ _____ _____

Comments on genetic results: _____

9. 12 Month Follow-Up

9.1 Follow-up

Follow-up data obtained
If yes, date

No Yes
|_|_|_| |_|_|_| |_|_|_|_|_|_|

Type of follow-up

Clinical Visit
 Telephone contact with Patient
 Telephone contact with family
 Hospital record
 Other

If other, please specify

Vital Status

If dead, date of death

Alive Dead
|_|_|_| |_|_|_| |_|_|_|_|_|_|

Cause(s) of death
Cardiovascular:

Procedure-related
 Non-procedure-related
 Unknown

Mode

Sudden Non-sudden

Cause

Acute myocardial infarction
 Heart failure
 Arrhythmia
 Ischaemic stroke
 Haemorrhagic stroke
 Systemic haemorrhage
 Peripheral embolism
 Pulmonary embolism
 Other

If other cardiac causes, please specify

Post-mortem examination

If yes,

No Yes
 Normal
 Inconclusive
 Diagnostic

Further details of histology: _____

9.2 Physical Characteristics

Height _____ cm

Weight _____ kg

Body mass index _____ Automatically calculated

Body surface area _____ Automatically calculated

9.3 Hospital Admission

<p>Has the patient been admitted to hospital since inclusion in the registry?</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes</p>
<p>Admission #1 If yes, date Reason for admission</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes</p> <p> _ _ _ _ _ _ _ _ </p> <p><input type="checkbox"/> Urgent admission for cardiac reason <input type="checkbox"/> Urgent admission for non cardiac reason <input type="checkbox"/> Planned admission for cardiac reason <input type="checkbox"/> Other</p>
<p>Admission #2 If yes, date Reason for admission</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes</p> <p> _ _ _ _ _ _ _ _ </p> <p><input type="checkbox"/> Urgent admission for cardiac reason <input type="checkbox"/> Urgent admission for non cardiac reason <input type="checkbox"/> Planned admission for cardiac reason <input type="checkbox"/> Other</p>
<p>Admission #3 If yes, date Reason for admission</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes</p> <p> _ _ _ _ _ _ _ _ </p> <p><input type="checkbox"/> Urgent admission for cardiac reason <input type="checkbox"/> Urgent admission for non cardiac reason <input type="checkbox"/> Planned admission for cardiac reason <input type="checkbox"/> Other</p>

<p>Admission #4</p> <p>If yes,</p> <p>date</p> <p>Reason for admission</p>	<p><input type="checkbox"/> No <input type="checkbox"/> Yes</p> <p> _ _ _ _ _ _ _ _ _ </p> <p><input type="checkbox"/> Urgent admission for cardiac reason</p> <p><input type="checkbox"/> Urgent admission for non cardiac reason</p> <p><input type="checkbox"/> Planned admission for cardiac reason</p> <p><input type="checkbox"/> Other</p>
--	--

9.4 Symptoms since inclusion

Syncope

- None
- Vagal or non-cardiac syncope
- Suspected arrhythmic/cardiogenic syncope
- Mechanism uncertain
- Not evaluated

Number of suspected _____
arrhythmic/cardiogenic syncope

Age at last |_|_| years
arrhythmic/cardiogenic syncope

Anginal chest pain No Yes

NYHA functional class NYHA I NYHA II NYHA III
 NYHA IV Not available/possible

Extreme lethargy No Yes

Palpitations No Yes

Ankle oedema No Yes

Orthopnea No Yes

Paroxysmal nocturnal dyspnoea No Yes

9.5 Arrhythmia since inclusion

Atrial Fibrillation No Yes

If yes, type of AF Paroxysmal Persistent

Permanent Unknown

Sustained ventricular tachycardia No Yes

Resuscitated ventricular fib./cardiac arrest No Yes

Stroke No TIA
 Stroke Not recorded

AV block No Yes

BBB No Yes

9.6 Investigations since inclusion

9.6.1 Electrocardiogram

ECG

Performed Not performed

Date (dd/mm/yyyy)

|_|_| | |_|_| | |_|_|_|_|_|

Rhythm

Sinus rhythm Atrial fibrillation
 Atrial flutter Atrial paced
 AV paced SR V paced
 AF V paced Ventricular tachycardia
 Other Not evaluated

Frequent premature beats No Yes

Heart rate _____ bpm

QT interval _____ msec

PR interval _____ ms

QRS duration _____ ms

QRS axis _____ degrees

Atrioventricular block No 1st degree

2nd degree 3rd degree

Bundle branch block No Incomplete RBBB

RBBB Incomplete LBBB

LBBB Other

Negative T waves No Yes Unknown

ST depression No Yes Unknown

ST elevation No Yes Unknown
Maximum R in praecordial _____ mm
Maximum S in praecordial _____ mm
Maximum R in limbs _____ mm
Maximum S in limbs _____ mm
Preexcitation No Yes
Abn Q-waves No Yes

9.6.2 Echocardiogram since inclusion

Echocardiogram Performed Not performed
Date (dd/mm/yyyy) |__|__| |__|__| |__|__|__|__|
LVEDD _____ mm
 _____ mm (*Automatically calculated*)
 Estimated LVEDD⁷ (Henry formula)
 _____ % (*Automatically calculated*)
 % of the estimated LV enddiastolic diameter⁸ (Henry formula)
LVESD _____ mm
 _____ %
LV ejection fraction (Simpson's biplane)
Fractional shortening _____ %
LVEDV _____ ml
LVESV (LV end systolic volume) _____ ml
Maximum LV thickness _____ mm
LV septal thickness diastole _____ mm
LV posterior wall thickness diastole _____ mm
Left atrium diameter _____ mm
Left atrial area _____ cm²
 Pericardial effusion No Yes

⁷ LVEDD calculated as follows: $((45,3 * ([BSA]^{(1/3)})) - (0,03 * [Age]) - 7,2)$

⁸ Calculated as follows: $([LVEDd]/[LVEDdEstHenry]) * 100$

Endocavitary thrombi

No Yes

Pattern of LV hypertrophy

No hypertrophy Asymmetrical septal

Concentric Apical

Other/not classified Not evaluated

RV dilation

No Yes

RV global systolic dysfunction

No Yes

RV hypertrophy

No Yes

Maximum RV wall thickness

_____ mm

New wall motion abnormality

No Yes

9.6.3 Doppler Diastolic Function Assessment since inclusion

Doppler performed

Performed Not performed

If yes, date

|_|_|_| |_|_|_| |_|_|_|_|_|_|

(dd/mm/yyyy)

Mitral E-wave

_____ m per second

E-wave deceleration time

_____ m per second

Mitral A-wave

_____ m per second

TDI lateral mitral annulus peak E velocity m/s (E or Ea)

_____ m per second

E/Ea (automatic calculation required to generate E/Ea (ie. Mitral E-wave m/s divided by Ea m/s)

Diastolic dysfunction (grade)

Normal
 Grade I (impaired relaxation)
 Grade II (pseudonormal)
 Restrictive
 Not evaluated

Left ventricular outflow tract gradient (resting)

_____ mmHg

Maximum provoked (by any technique) peak left ventricular outflow tract gradient

_____ mmHg

Aortic regurgitation

None Mild
 Moderate Severe

Mitral regurgitation

None Mild
 Moderate Severe

Tricuspid regurgitation

None Mild
 Moderate Severe

Systolic Pulmonary Artery pressure

_____ mmHg

Not possible/not evaluated

Aortic stenosis

None Mild

Moderate Severe

Mitral stenosis

None Mild

Moderate Severe

Valve replacement

No Yes

If yes, replaced valves

Mitral Aortic Tricuspid Pulmonary

9.6.4 Cardiac Magnetic Resonance Imaging since inclusion

MRI scan

Not performed Normal

Abnormal Inconclusive

If performed, date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

Late gadolinium enhancement (LGE)

No Yes

Not possible/not evaluated

LGE localisation, specify

LV end-diastolic volume

_____ ml

LV end-systolic volume

_____ ml

LV ejection fraction

_____ % (*Absolute number*)

Maximum LV thickness by CMR

_____ mm

Left ventricular mass (LVM)

_____ g

Pattern of hypertrophy

No hypertrophy Septal

Concentric Eccentric

Apical Other

Unknown

9.6.5 Holter since inclusion

Holter performed

Performed Not performed

If yes, date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Non-sustained ventricular tachycardia

No Yes Unknown

Rhythm

- Sinus rhythm throughout
- Paroxysmal atrial fibrillation in sinus rhythm
- Atrial fibrillation throughout
- Sinus rhythm & V pacing
- AV pacing
- Permanent atrial fibrillation & V pacing
- Conduction defect (AVB 1-2-3; sinus dysfunction)

9.6.6 Exercise Test since inclusion

Exercise test performed

Performed Not performed

If yes, date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Exercise type

Bicycle Treadmill

Medication on day of exercise test

- No
- Beta-blocker
- Verapamil or Diltiazem
- Other
- Unknown

If Other, please specify

Systolic blood pressure, baseline

_____ mmHg

Systolic blood pressure, max achieved

_____ mmHg

Systolic blood pressure, peak exercise

_____ mmHg

Absolute workload achieved

_____ Watts

_____ METS

Peak heart rate

_____ bpm

Ventricular arrhythmia

No Yes

Supraventricular arrhythmia

No

Yes

9.6.7 Laboratory since inclusion

Collection performed

No

Yes

If yes, please complete below

Collection date (dd/mm/yyyy)

|_|_|_| |_|_|_| |_|_|_|_|_|_|

Haemoglobin

_____ mmol/L g/dl

White blood cells

_____ $10^3/\text{mm}^3$ $10^9/\text{L}$

Platelets

_____ $10^3/\text{mm}^3$ $10^9/\text{L}$

Creatine kinase

_____ IU/L

Troponin I

_____ ng/ml

Troponin T

_____ ng/ml

Creatinine

_____ $\mu\text{mol}/\text{L}$ mg/dl

Total cholesterol

_____ mmol/L mg/dL

Fasting glucose

_____ mmol/L mg/dL

Sodium

_____ mEq/L mmol/L

Potassium

_____ mEq/L mmol/L

Uric acid

_____ $\mu\text{mol}/\text{L}$ mg/dL

HbA1c

_____ % mmol/mol

Nitrogen measured by:

BUN (*Blood Urea Nitrogen*)

_____ mmol/L mg/dL

Urea

_____ mmol/L mg/dL

BNP

_____ pmol/L pg/mL

NT-proBNP

_____ pmol/L pg/mL

T4

_____ mEq/L mmol/L

pmol/L ng/dL

TSH

_____ mIU/L

9.6.7 Signal averaged ECG

Date of Signal averaged ECG:

- Normal (0/3) 1/3 criteria
- 2/3 criteria 3/3 criteria
- Not performed

9.7 Medications

Is the patient on any medication? No Yes

Alpha-blockers	<input type="checkbox"/> No <input type="checkbox"/> Yes
Beta-blockers	<input type="checkbox"/> No <input type="checkbox"/> Sotalol <input type="checkbox"/> Carvedilol <input type="checkbox"/> Bisoprolol <input type="checkbox"/> Metoprolol <input type="checkbox"/> Nebivolol <input type="checkbox"/> Other If other, please specify: _____ Dose: _____
Diuretics, oral <i>(Multiple possible answers)</i>	<input type="checkbox"/> No <input type="checkbox"/> Bendrofluazide <input type="checkbox"/> Hydrochlorothiazide <input type="checkbox"/> Indapamide <input type="checkbox"/> Torasemide <input type="checkbox"/> Chlorthalidone <input type="checkbox"/> Furosemide <input type="checkbox"/> Bumetanide <input type="checkbox"/> Other If other, please specify: _____ Dose: ____
ACE inhibitors	<input type="checkbox"/> No <input type="checkbox"/> Perindopril <input type="checkbox"/> Ramipril <input type="checkbox"/> Lisinopril <input type="checkbox"/> Enalapril <input type="checkbox"/> Captopril <input type="checkbox"/> Fosinopril <input type="checkbox"/> Other If other, please specify: _____ Dose: _____
Angiotensin II receptor blockers	<input type="checkbox"/> No <input type="checkbox"/> Candesartan <input type="checkbox"/> Losartan <input type="checkbox"/> Valsartan <input type="checkbox"/> Other If other, please specify: _____ Dose: _____
	If other, please specify: _____ Dose: _____
Mineralocorticoid antagonists (MRAs)	<input type="checkbox"/> No <input type="checkbox"/> Spironolactone <input type="checkbox"/> Eplerenone <input type="checkbox"/> Canrenone <input type="checkbox"/> Other If other, please specify: _____ Dose: _____

Antiplatelets	<input type="checkbox"/> No <input type="checkbox"/> Clopidogrel <input type="checkbox"/> Prasugrel <input type="checkbox"/> Ticagrelor <input type="checkbox"/> Aspirin <input type="checkbox"/> Other If other, please specify: _____ Dose: _____
Oral anticoagulants	<input type="checkbox"/> No <input type="checkbox"/> Vit K Antagonists <input type="checkbox"/> Dabigatran <input type="checkbox"/> Rivaroxaban <input type="checkbox"/> Apixaban <input type="checkbox"/> Other If other, please specify: _____ Dose: _____
Anti-arrhythmic drugs <i>(Multiple possible answers)</i>	<input type="checkbox"/> No <input type="checkbox"/> Amiodarone <input type="checkbox"/> Digitoxin/digoxin <input type="checkbox"/> Flecainide <input type="checkbox"/> Propafenone <input type="checkbox"/> Disopyramide <input type="checkbox"/> Other If other, please specify: _____
Calcium antagonists	<input type="checkbox"/> No <input type="checkbox"/> Diltiazem <input type="checkbox"/> Verapamil <input type="checkbox"/> Other: _____ Dose: _____
Nitrates	<input type="checkbox"/> No <input type="checkbox"/> Yes
Statins	<input type="checkbox"/> No <input type="checkbox"/> Yes
Ivabradine	<input type="checkbox"/> No <input type="checkbox"/> Yes
Direct renin inhibitors	<input type="checkbox"/> No <input type="checkbox"/> Yes
Other drugs, specify: _____	

Comments on medication: _____

9.9 Device Therapy since last visit

(Tick the boxes ✓ or fill in the spaces)

New or upgraded device since last visit

No Yes

Generator change since last visit

No Yes

Cardioverter defibrillator implanted

No Yes

If yes, date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Reason for cardioverter defibrillator

Yes, Primary prophylaxis

Yes, Secondary prophylaxis

Pacemaker implanted

No Yes

If yes, date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Reason for pacemaker

(Multiple answers possible)

Bradyarrhythmia

Treatment of left ventricular outflow tract obstruction

Cardiac resynchronisation therapy

Other

9.10 Other Procedures since last visit

Septal myectomy

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Alcohol septal ablation

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Heart transplant

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Ventricular assist device

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

EP study performed

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

Cardiac ablation

No Yes

If yes

For ventricular tachycardia

No Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|_|

For atrial fibrillation

No

Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|

For nodal tachycardia

No

Yes

Date (dd/mm/yyyy)

|_|_| |_|_| |_|_|_|_|
