

The Echocardiographic Assessment of the Right Ventricle with particular reference to Arrhythmogenic Right Ventricular Cardiomyopathy – A Protocol of the British Society of Echocardiography

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### **Preamble**

Assessment of the right ventricle (RV) is often challenging and sometimes overlooked, however recent guideline documentation from the American Society of Echocardiography suggested a measure of RV structure and function should be mandatory in all clinical reports\*. The BSE advocates RV assessment within the minimum dataset; however in certain conditions such as arrhythmogenic right ventricular cardiomyopathy (ARVC), pulmonary hypertension, pulmonary embolism, RV myocardial infarction and athletic heart syndrome a more comprehensive assessment of the RV is required. RV assessment can be described in terms of RV dimensions, structure and function and the assessment of ARVC utilises this approach. It is clear that with other RV pathology the measurements are similar but their interpretation should be taken in the clinical context.

ARVC is one of the most common and under-diagnosed causes of cardiac sudden death in a young person and therefore an appropriate diagnosis is crucial. Echocardiography has variable sensitivity and specificity for the diagnosis of ARVC and therefore only forms a small part of the complete diagnosis. Corroborative investigations are key and include a comprehensive history, clinical examination, electrocardiogram, magnetic resonance imaging and genetic testing all contributing to the overall assessment. Echocardiographic criteria demonstrated in isolation should be interpreted with caution and therefore although this document is a protocol for RV assessment *per se*, it should be used only as part of the assessment for ARVC.

Table 1- Echocardiographic criteria for ARVC (adapted from Marcus et al 2010)

# MAJOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC

Regional RV Dyskinesia or Aneurysm

And one of the following

PLAX RVOT  $\geq$  32mm (corrected for body size [PLAX/BSA]  $\geq$  19mm/m2)

PSAX RVOT  $\geq$  36mm (corrected for body size [PLAX/BSA]  $\geq$  21mm/m2)

Or

Fractional Area Change ≤ 33%

## MINOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC

Regional RV Akinesia or Dyskinesia

And one of the following

PLAX RVOT  $\geq$  29 to < 32mm (corrected for body size [PLAX/BSA]  $\geq$  16 to < 19mm/m2)

PSAX RVOT ≥ 32 to < 36mm (corrected for body size [PLAX/BSA] ≥ 18 to 21mm/m2)

Or

Fractional Area Change > 33 to < 40%

VIEW	Modality	Measurements	Explanatory note for ARVC	Image
PLAX	2D	RVOT <sub>PLAX</sub> Qualitative regional wall motion analysis of the anterior wall of the RV	-end diastole* -adjust depth and focal zone to visualise RVOTfor consistency, ideally, this measurement should be taken at a similar level to RVOT₁ measurement of PSAX AV view. Hence RVOT₽LAX should be a measurement perpendicular line from the RV anterior wall to the level of the aortic valveall 2D measurements should be blood tissue interface to blood tissue interface RVOT₽LAX ≥ 32mm or ≥ 19mm/m² AND the presence of regional RV akinesia, dyskinesia or aneurysm is a major criterion**  RVOT₽LAX ≥ 29mm to < 32mm OR ≥ 16mm/m² to <19mm/m² AND the presence of regional RV akinesia or dyskinesia is a minor criterion**	OX1 S5-1 36Hz 14cm HiGen Gn 60 C3 72 / 0 75 mm/s  AV P A R R R R R R R R R R R R R R R R R R
PLAX RV inflow	2D	Qualitative regional wall motion analysis of the anterior and inferi- or walls of the RV	-ensure the ventricular septum has been excluded and the true inferior wall is seen (diaphragm and liver in view)	DXI 85-1 100Hz 13-4cm Gn 60 C 50 3/2/0 75 mm/s  Anterior Wall
PLAX RV inflow	Colour Flow Doppler	Assess the severity of tricuspid regurgitation and estimate RV systolic pressure (for details see pulmonary hypertension dataset)	The presence of TR is not a sensitive or specific finding for ARVC however severe functional TR may occur in the presence of RV dilatation and dysfunction	Tricuspid  Regurgitation    Tricuspid   Regurgitation   Regurg
PSAX AV level	2D	Proximal RVOT (RVOT <sub>1</sub> )  Qualitative assessment of RV structure and function  Regional wall motion analysis of the outflow tract of the RV (infundibulum)	-at end diastole* -measured from anterior aortic wall directly up to the RV free wall (at the level of the aortic valve) -the PSAX view has been shown to be more reproducible than the measurement obtained from the PLAX orientation  RVOT1 ≥ 36mm or ≥ 21mm/m² in the presence of regional RV akinesia, dyskinesia or aneurysm is a major criterion**	37.1 S5-1 S7-1 S7-1 S7-1 S7-1 S7-1 S7-1 S7-1 S7

			RVOT1 ≥ 32mm to < 36mm or ≥ 18mm/m² to <21mm/m² in the presence of regional RV akinesia or dyskinesia is a minor criteri- on**	
PSAX PV level	2D	Distal RVOT (RVOT <sub>2</sub> )  Qualitative assessment of RV structure and function  Regional wall motion analysis of the infundibulum of the RV  PA diameter	-end diastole* -measured just proximal to PV  There are no specific values for diagnosis of ARVC however this should be used to demonstrate dilatation.  RVOT <sub>2</sub> > 27mm is abnormal in other cardiac pathology*  -end diastole - half way between pulmonary valve (PV) and bifurcation of main PA or 1cm distal to PV Enlargement of the pulmonary artery makes the diagnosis of ARVC less likely (may be indicative of conditions causing pulmonary hypertension)	OX1
PSAX Base	2D	Qualitative assessment of RV structure and function at basal level  Regional wall motion analysis of inferior, lateral, anterior and septal walls of RV in PSAX at base (mitral valve) level	Relative size of RV to LV should be assessed  There is disproportionate enlargement of the RV in ARVC	DAVE OXBOI SS 1 SS
PSAX Mid	2D	Qualitative assessment of RV structure and function at papillary muscle level  Regional wall motion analysis of inferior, lateral, anterior and septal walls of RV in PSAX at mid (papillary muscle) level	Relative size of RV to LV should be assessed	DAVE OXBOI SS-1 SS-1 SS-1 SS-1 RV Lateral Wall RV Anterior Wall 11cm 2D HGen Gn 36 C 39
PSAX Apex		Qualitative assessment of RV structure and function at the apex Regional wall motion analysis of inferior, lat- eral and septal walls of RV in PSAX at apex level	Relative size of RV to LV should be assessed	DAVE OXBOI SS-1 SS-1 SS-1 SS-1 SS-1 SS-1 SS-1 SS-

Apical 4CH Focused RV view	2D	RVD <sub>1</sub> – Basal RV diameter (end diastole at the maximal value within the first third of the RV)*  RVD <sub>2</sub> – Mid RV diameter (end diastole in the middle third of the RV at the level of the LV papillary muscles)  RVD <sub>3</sub> – RV length (end diastole from tricuspid annulus to the RV apex)  Fractional Area Change (FAC)  Qualitative assessment of RV structure and longitudinal function	Focused RV 4CH view is obtained by ensuring:  1. true apex is visualised, with scan plane positioned through the LV in the centre of the cavity 2. RV is not foreshortened and LVOT is not opened  3. largest RV dimensions are optimised while maintaining 'on axis' view, as described above (for further clarification see ASE RV guidelines*)  There are no specific values for diagnosis of ARVC however all RV measurements should be used to demonstrate dilatation. RVD <sub>1</sub> > 42mm, RVD <sub>2</sub> > 35mm and RVD <sub>3</sub> > 86mm are abnormal*  -trace around the endocardium of the RV lateral wall at end diastole and end systoledo not trace around individual trabeculations, which should be included within the cavity area.)	RVD2  RVD1  RVD1  RVD1  RVD2  RVD2
			FAC ≤ 33% in the presence of regional RV akinesia, dyskinesia or aneurysm is a major criterion** even in the presence of normal RVOT size.  FAC > 33% to ≤ 40%in the presence of regional RV akinesia or dyskinesia is a minor criterion** even in the presence of normal RVOT size.	RV Systolic Area
АР4СН	M-Mode	Tricuspid Plane Systolic Excursion (TAPSE)	Ensure correct alignment of RV, such that RV base moves perpendicular to scan plane and is not oblique. The latter will cause a falsely reduced TAPSE value  There are no specific values for diagnosis of ARVC however TAPSE should be used to demonstrate longitudinal dysfunction. TAPSE < 16mm is abnormal*	0X1
AP4CH	PW Doppler	E and A wave peak velocities for RV dias- tolic function using trans-tricuspid PW Doppler (optional)	There are no specific values for diagnosis of ARVC however diastolic dysfunction may indicate early changes in overall RV function. E < 0.35cm/s and E:A ratio < 0.8 may indicate impairment in diastolic filling*	OX1 + Vel 51.0 cm/s SS-1 PG 1.17 cm/s SS-1 PG 1.17 cm/s 17 cm PG 0.305 cm/s 18 cm PG 0.305 cm/s 19 cm PG 0.305 cm/s 10 cm PG 0
	Tissue Doppler	Systolic (S'), early (E') and atrial (A') relaxation velocities at lateral TV annulus	There are no specific values for diagnosis of ARVC however TDI should be used to demonstrate longitudinal systolic and/or diastolic dysfunction. s' < 10cm/s, e' < 8cm/s and A' < 7cm/s are abnormal* .An E/e' of > 6 may be consistent with an elevated RA pressure.	# 15 / 0   1   1   1   1   1   1   1   1   1

Modified AP4CH (medi- al movement of the angle of the ultra- sound beam)	Colour Flow Doppler	Assess the severity of Tricuspid Regurgitation and estimate RV sys- tolic pressure		10. 37  15. 37  16. 32  17. TX Procx 2.4 m/n 17. TX maxPG 23.6 mmHz 15. 37  16. 37  17. 18. 18. 18. 18. 18. 18. 18. 18. 18. 18
Useful additional parameters standard Apical 4CH	2D	Basal RV:LV ratio at end diastole.	There are no specific values for diagnosis of ARVC however the measurement may be used to demonstrate RV dilatation. RV:LV ratio > 0.66 is abnormal*	0X1 83-1 31Hz 16cm Gn 60 C 30 / 75 mm/s RV5.5cm LV 4.5cm
		Qualitative assessment of RV structure and longitudinal function.  Detection of regional RV dyskinesia or aneurysm formation is part of the major echocardiographic criteria for ARVC	A thickened or echo-bright moderator band is not specific for ARVC but may support the diagnosis in the presence of other findings	FR 45Hz  TRV Aneurysm - Thin  Wall, Thickened Moderator Band  FR 45Hz  Tem  As bpm  FR 45Hz  Tem  As bpm  FR 45Hz  Tem  As aneurysms, thin wall  Tem  As aneurysms, thin wall  Tem  Tem  Tem  Tem  Tem  Tem  Tem  T
		RA area at ventricular end systole	There are no specific values for diagnosis of ARVC however the measurement should be used to demonstrate RA dilatation. RA area > 18cm² is abnormal*	Dilated Right Atrium

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Apical 5CH	2D	Identify thickened moderator band	Outflow tract of the RV ( infundibulum) /thickened moderator band is not specific for ARVC but may support the diagnosis in the presence of other findings	Thickened Moderator Band  10
Sub-costal	2D	Qualitative assessment of RV structure and function	Regional wall motion analysis of inferior wall of RV	RV Aneurysm
		RV wall thickness	- at end diastole - ignore trabeculations and papillary muscles - use reduced depth to improve resolution and measurement accuracy  There are no specific values for diagnosis of ARVC however the measurement should be used to demonstrate RV thinning <3mm. RV wall thickness > 5mm is consistent with RV hypertrophy.*	0X1
		IVC size and inspiratory collapse	Estimate of RA pressure to define RV end systolic pressure (see pulmonary hypertension protocol for details)	OX1  OX1  OX1  OX1  OX1  OX1  OX1  OX1
				\$5-1 \$4ttz

Sub-costal	Colour Flow Doppler	Assess the severity of Tricuspid Regurgitation and estimate RV sys- tolic pressure	The presence of TR is not a sensitive or specific finding for ARVC however significant functional TR may occur in the presence of RV dilatation and dysfunction	OX1 S3-1 13HZ 16EM 16EM 16EM 17
	CW		May perform if good Doppler alignment of	1 TE Vmox 2.4 m/s TR moxPG 22.8 mmHg 15.  0.353  (m/s) 15.  1.0  1.1.0
	Doppler		Tricuspid Regurgitation jet direction	

### ADDITIONAL NOTES

- These values should be interpreted with caution in the athletic population;
- RV akinesia, dyskinesia or aneurysm are diagnostic criteria in the presence of RV dilatation or reduced RV fractional area change\*\*
- Assess the LV in line with the BSE minimum dataset LV involvement may occur early in the course of the disease†
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- † Sen-Chowdhry S, Syrris P, Prasad SK, Hughes SE, Merrifield R, Ward D, Pennell DJ, McKenna WJ. Left-dominant arrhythmogenic cardiomyopathy: an under-recognized clinical entity. *J Am Coll Cardiol.* 2008;52:2175–2187.
- ‡ Oxborough D, Sharma S, Shave R, Whyte G, Birch K, Artis N, Batterham A, George K The right ventricle of the endurance athlete: the relationship between morphology and deformation. *J Am Soc Echocardiogr* 25(3):263-271