British Society of Echocardiography

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

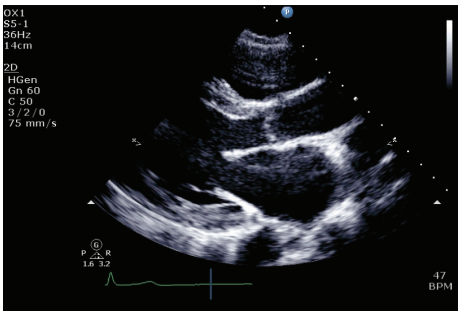
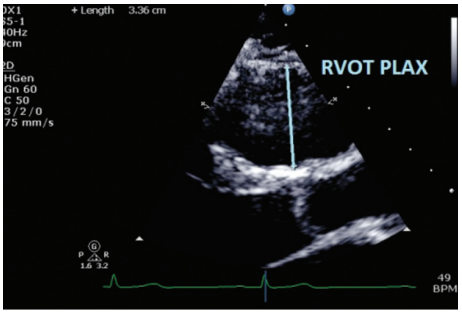
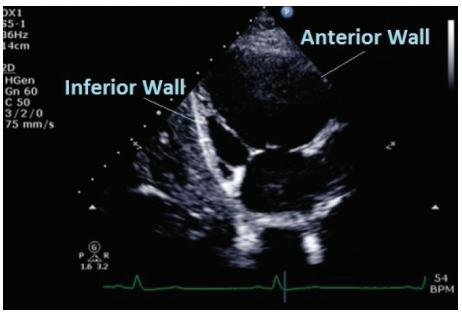
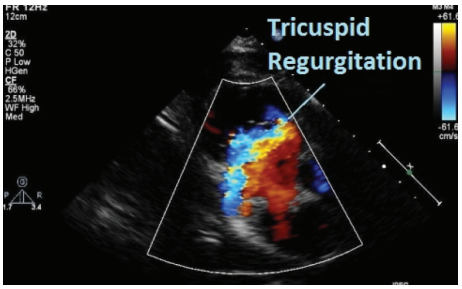
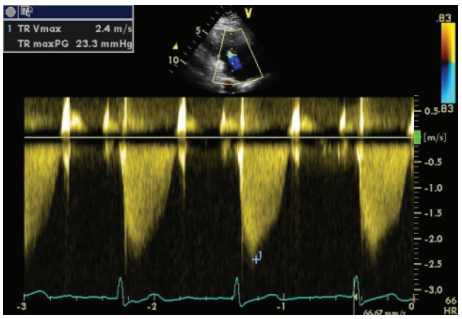
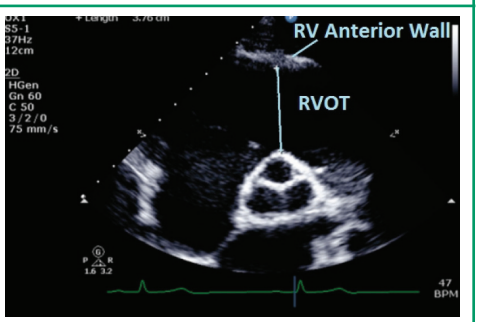
Assessment of the right ventricle (RV) is often challenging and sometimes overlooked, however recent guideline documentation from the American Society of Echocardiography suggested a measure of RV structure and function should be mandatory in all clinical reports*. The BSE advocates RV assessment within 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.

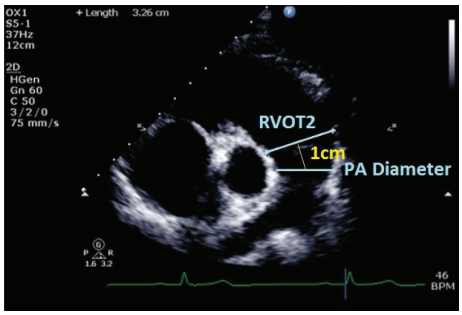
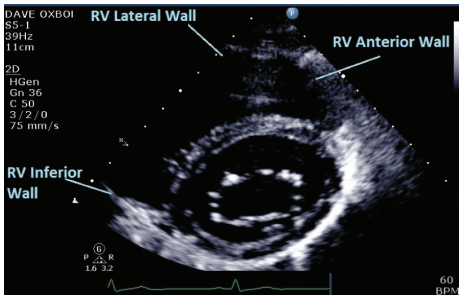
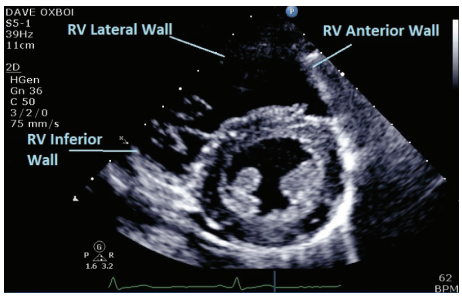
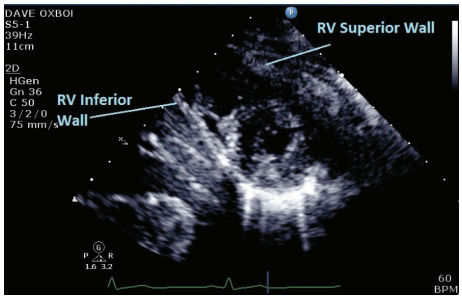
ARVC is one of the most common and under-diagnosed causes of cardiac sudden death in a young person and therefore an appropriate diagnosis is crucial. Echocardiography has variable sensitivity and specificity for the diagnosis of ARVC and therefore only forms a small part of the complete diagnosis. Corroborative 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.

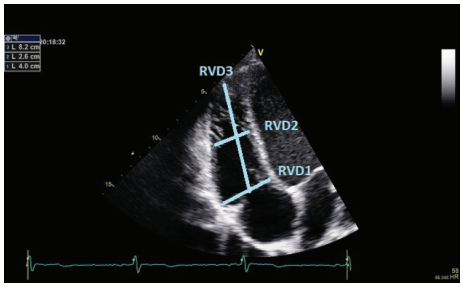
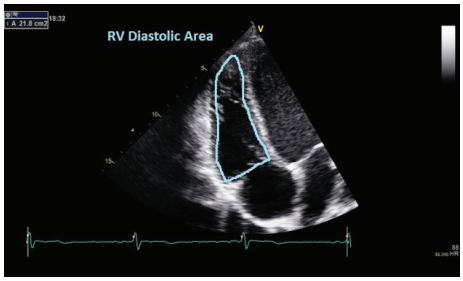
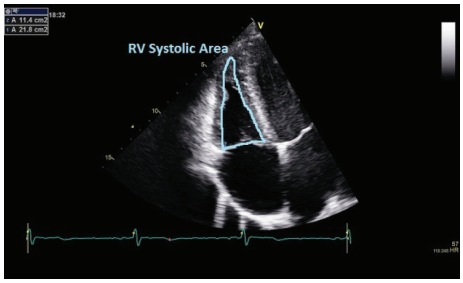
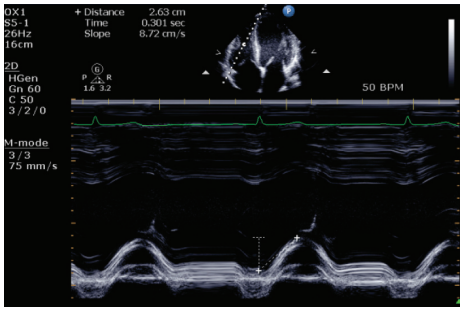
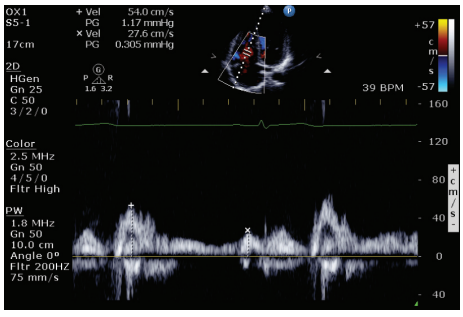
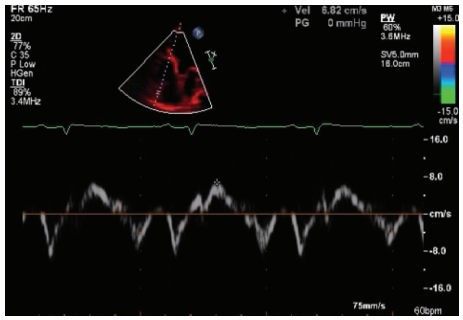
Table 1- Echocardiographic criteria for ARVC (adapted from Marcus et al 2010)

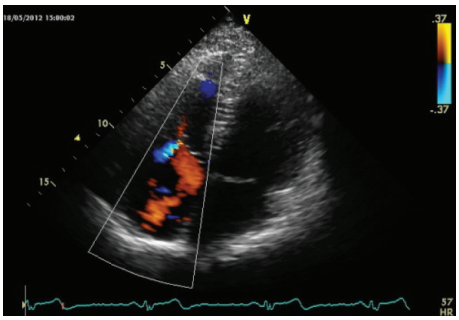
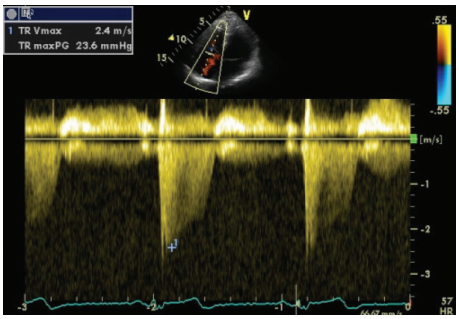
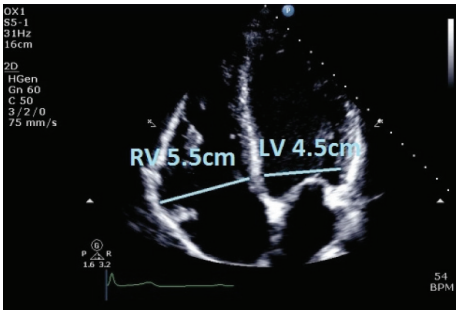
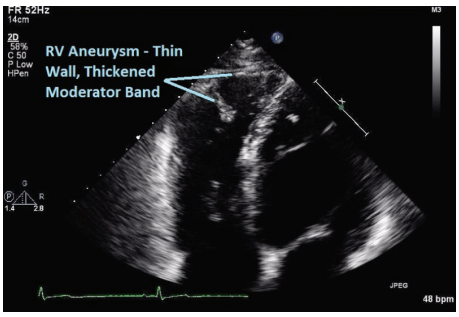
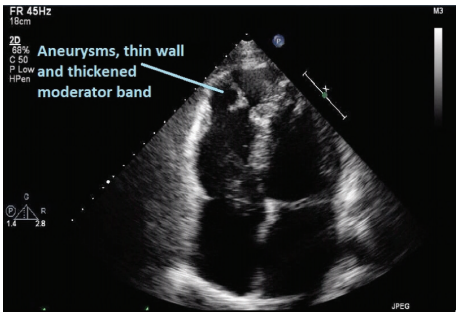
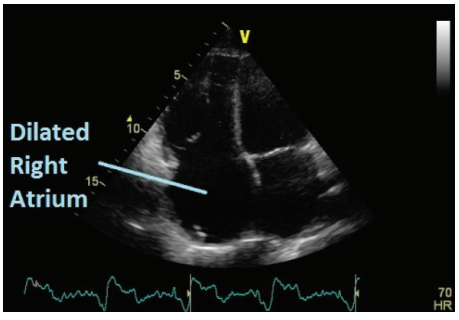
MAJOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC
Regional RV Dyskinesia or Aneurysm
And one of the following
PLAX RVOT ≥ 32mm (corrected for body size [PLAX/BSA] ≥ 19mm/m2)
PSAX RVOT ≥ 36mm (corrected for body size [PLAX/BSA] ≥ 21mm/m2)
Or
Fractional Area Change ≤ 33%

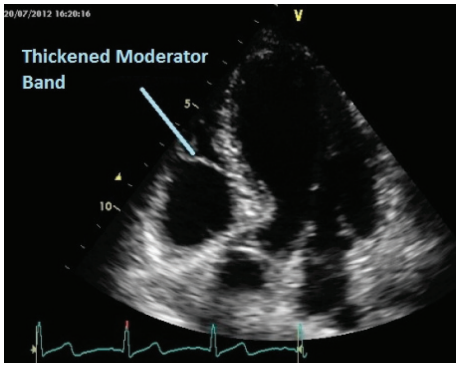
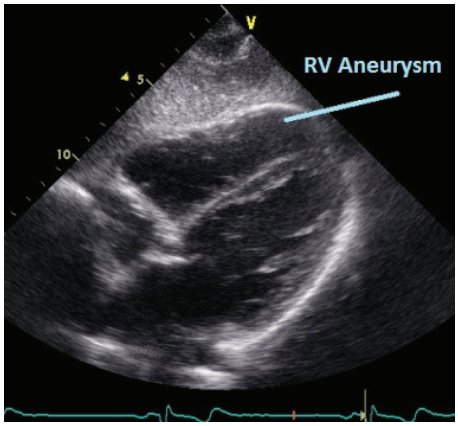
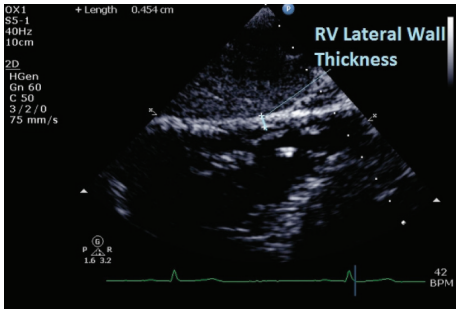
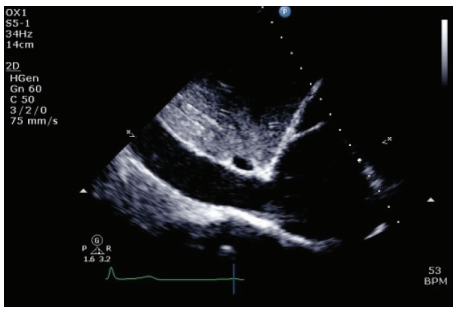
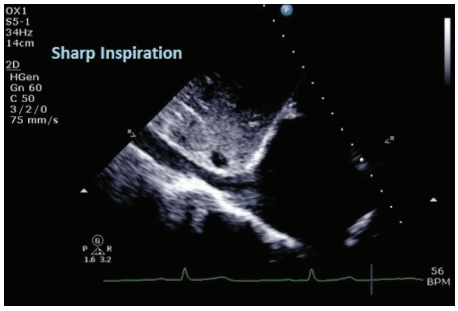
MINOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC
Regional RV Akinesia or Dyskinesia
And one of the following
PLAX RVOT ≥ 29 to < 32mm (corrected for body size [PLAX/BSA] ≥ 16 to < 19mm/m2)
PSAX RVOT ≥ 32 to < 36mm (corrected for body size [PLAX/BSA] ≥ 18 to 21mm/m2)
Or
Fractional Area Change > 33 to < 40%

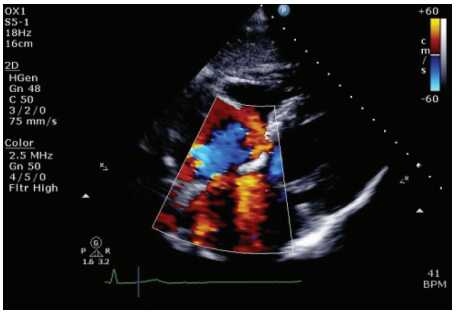
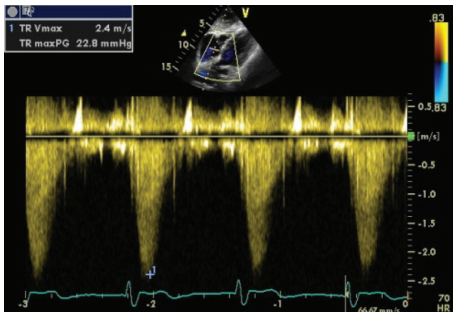
VIEW	Modality	Measurements	Explanatory note for ARVC	Image
PLAX	2D	**$RVOT_{PLAX}$** Qualitative regional wall motion analysis of the anterior wall of the RV	-end diastole* -adjust depth and focal zone to visualise RVOT. -for consistency, ideally, this measurement should be taken at a similar level to $RVOT_1$ measurement of PSAX AV view. Hence $RVOT_{PLAX}$ should be a measurement perpendicular line from the RV anterior wall to the level of the aortic valve. -all 2D measurements should be blood tissue interface to blood tissue interface $RVOT_{PLAX} \geq$ 32mm or $\geq$ 19mm/m^2 AND the presence of regional RV akinesia, dyskinesia or aneurysm is a major criterion** $RVOT_{PLAX} \geq$ 29mm to < 32mm OR ≥ 16mm/m^2 to <19mm/m^2 AND the presence of regional RV akinesia or dyskinesia is a minor criterion**	 
PLAX RV inflow	2D	Qualitative regional wall motion analysis of the anterior and inferior walls of the RV	-ensure the ventricular septum has been excluded and the true inferior wall is seen (diaphragm and liver in view)	
PLAX RV inflow	Colour Flow Doppler CW 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	 
PSAX AV level	2D	**Proximal RVOT ($RVOT_1$)** 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 $\geq$ 36mm or $\geq$ 21mm/m^2 in the presence of regional RV akinesia, dyskinesia or aneurysm is a major criterion**	

			RVOT1 ≥ 32mm to < 36mm or ≥ 18mm/m^2 to <21mm/m^2 in the presence of regional RV akinesia or dyskinesia is a minor criterion**	
PSAX PV level	2D	Distal RVOT ($RVOT_2$) 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. $RVOT_2$ > 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)	
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	
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	
PSAX Apex		Qualitative assessment of RV structure and function at the apex Regional wall motion analysis of inferior, lateral and septal walls of RV in PSAX at apex level	Relative size of RV to LV should be assessed	

Apical 4CH Focused RV view	2D	**RVD_1** – Basal RV diameter (end diastole at the maximal value within the first third of the RV)* **RVD_2** – Mid RV diameter (end diastole in the middle third of the RV at the level of the LV papillary muscles) **RVD_3** – 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_1 > 42mm, RVD_2 > 35mm and RVD_3 > 86mm are abnormal* -trace around the endocardium of the RV lateral wall at end diastole and end systole. -do not trace around individual trabeculations, which should be included within the cavity area.) 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.	  
AP4CH	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*	
AP4CH	PW Doppler Tissue Doppler	**E and A wave peak velocities** for RV diastolic function using trans-tricuspid PW Doppler (optional) **Systolic (S'), early (E') and atrial (A') relaxation** velocities at lateral TV annulus	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* 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.	 

Modified AP4CH (medial movement of the angle of the ultrasound beam)	Colour Flow Doppler	Assess the severity of Tricuspid Regurgitation and estimate RV systolic pressure		
	CW Doppler			
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*	
		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	 
		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 > 18cm^2 is abnormal*	

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	
Sub-costal	2D	Qualitative assessment of RV structure and function	Regional wall motion analysis of inferior wall of RV	
		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.*	
		IVC size and inspiratory collapse	Estimate of RA pressure to define RV end systolic pressure (see pulmonary hypertension protocol for details)	 

Sub-costal	Colour Flow Doppler	Assess the severity of Tricuspid Regurgitation and estimate RV systolic 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	 
	CW Doppler		May perform if good Doppler alignment of 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†

* Rudski, L. G., Lai, W. W., Afilalo, J., Hua, L., Handschumacher, M. D., Chandrasekaran, K., Solomon, S. D., Louie, E. K. & Schiller, N. B. 2010. Guidelines for the Echocardiographic Assessment of the Right Heart in Adults: A Report from the American Society of Echocardiography: Endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography. *Journal of the American Society of Echocardiography*, 23, 685-713.

** Marcus, F. I., Mckenna, W. J., Sherrill, D., Basso, C., Bauce, B., Bluemke, D. A., Calkins, H., Corrado, D., Cox, M. G. P. J., Daubert, J. P., Fontaine, G., Gear, K., Hauer, R., Nava, A., Picard, M. H., Protonotarios, N., Saffitz, J. E., Sanborn, D. M. Y., Steinberg, J. S., Tandri, H., Thiene, G., Towbin, J. A., Tsatsopoulou, A., Wichter, T. & Zareba, W. 2010. Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia. *Circulation*, 121, 1533-1541.

† Sen-Chowdhry S, Syrris P, Prasad SK, Hughes SE, Merrifield R, Ward D, Pennell DJ, McKenna WJ. Left-dominant arrhythmogenic cardiomyopathy: an underrecognized 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