

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

Lead Authors

Dr. David Oxborough, Dr Abbas Zaidi, Prof Sanjay Sharma, Prof John Somauroo

Education Committee Authors

Dr Rick Steeds (Chair), Will Bradlow, Alison Carr, Richard Jones, Prathap Kanagala, Daniel Knight, Guy Lloyd, Thomas Mathew, Navroz Masani, Kevin O’Gallagher, Bushra Rana, Liam Ring, Julie Sandoval, Martin Stout, Gill Wharton, Richard Wheeler

Preamble

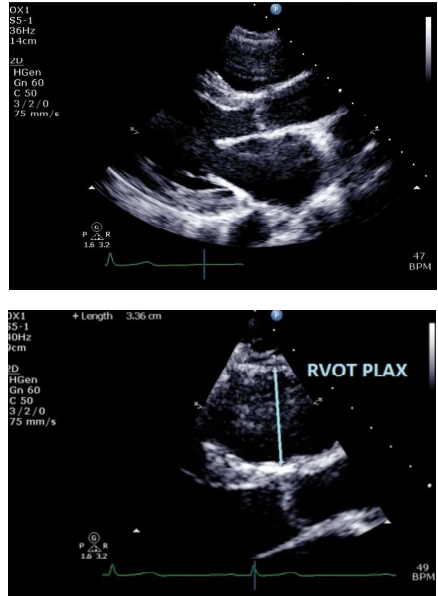
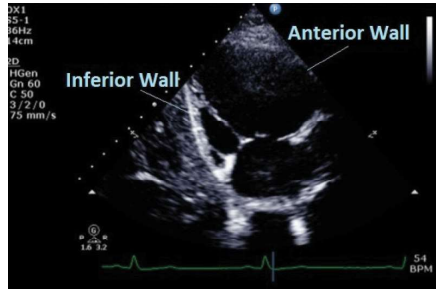
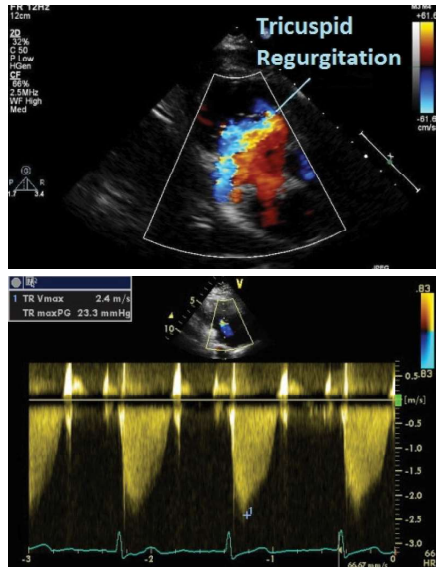
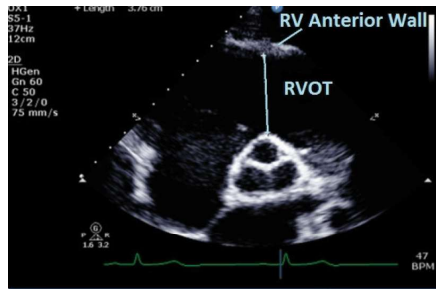
Assessment of the right ventricle (RV) is often challenging and sometimes overlooked, however recent guideline documentation from the American Society of Echocardiography suggested a measure of RV structure and function should be mandatory in all clinical reports*. The BSE advocates RV assessment within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

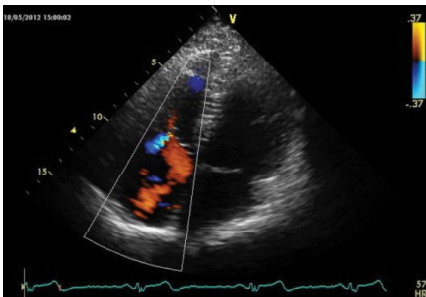
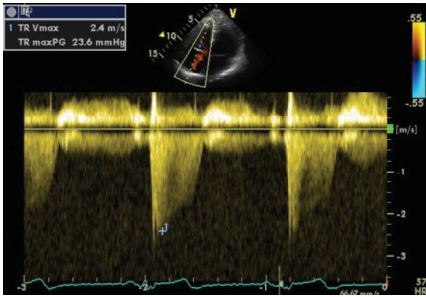
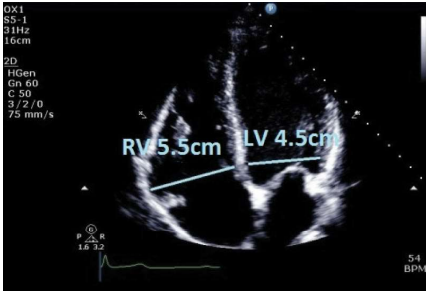
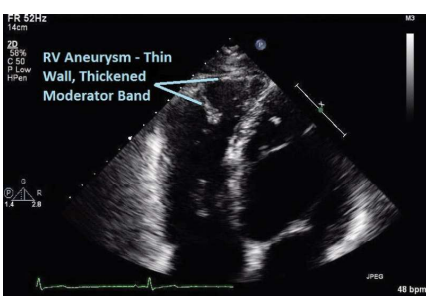
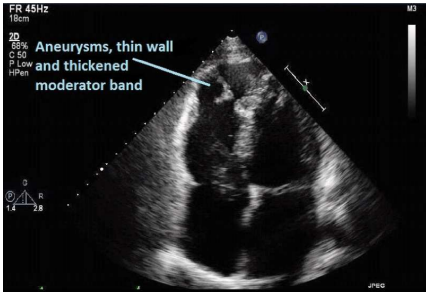
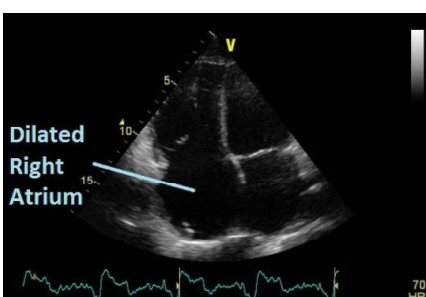
Table 1- Echocardiographic criteria for ARVC (adapted from Marcus et al 2010)

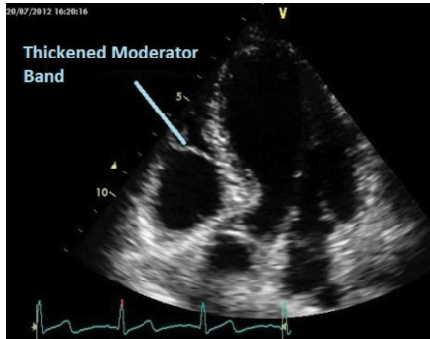
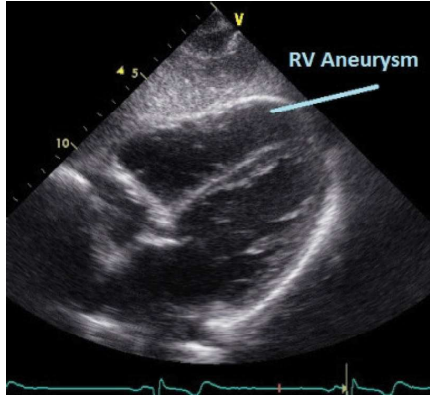
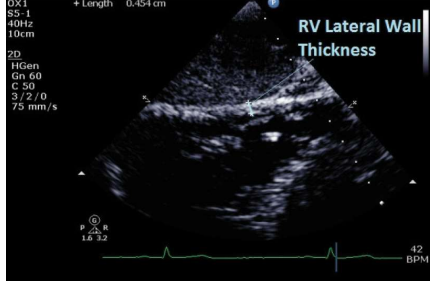
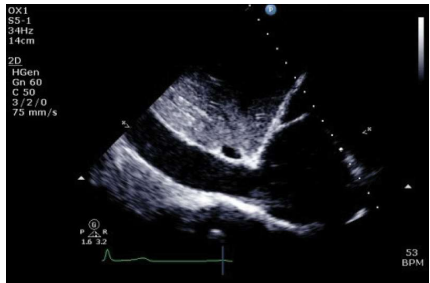
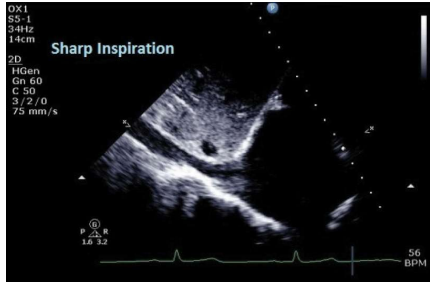
| MAJOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC |
|--|
| Regional RV Dyskinesia or Aneurysm |
| <i>And one of the following</i> |
| PLAX RVOT $\geq 32\text{mm}$ (corrected for body size $[\text{PLAX/BSA}] \geq 19\text{mm/m}^2$) |
| PSAX RVOT $\geq 36\text{mm}$ (corrected for body size $[\text{PLAX/BSA}] \geq 21\text{mm/m}^2$) |
| <i>Or</i> |
| Fractional Area Change $\leq 33\%$ |

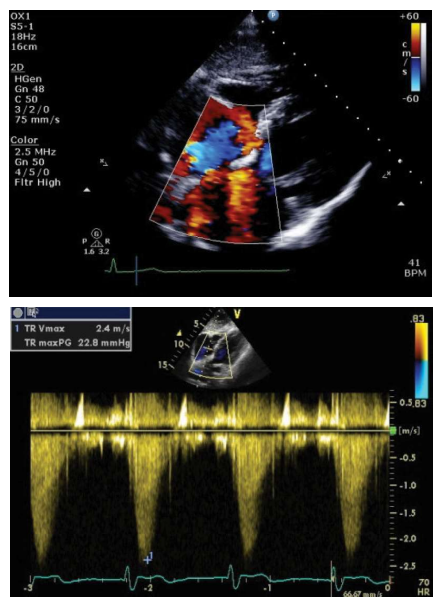
| MINOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC |
|--|
| Regional RV Akinesia or Dyskinesia |
| <i>And one of the following</i> |
| PLAX RVOT ≥ 29 to $< 32\text{mm}$ (corrected for body size $[\text{PLAX/BSA}] \geq 16$ to $< 19\text{mm/m}^2$) |
| PSAX RVOT ≥ 32 to $< 36\text{mm}$ (corrected for body size $[\text{PLAX/BSA}] \geq 18$ to 21mm/m^2) |
| <i>Or</i> |
| Fractional Area Change > 33 to $< 40\%$ |

| VIEW | Modality | Measurements | Explanatory note for ARVC | Image |
|----------------|---|--|---|---|
| PLAX | 2D | <p>RVOT_{PLAX}</p> <p>Qualitative regional wall motion analysis of the anterior wall of the RV</p> | <p>-end diastole*</p> <p>-adjust depth and focal zone to visualise RVOT.</p> <p>-for consistency, ideally, this measurement should be taken at a similar level to RVOT₁ 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.</p> <p>-all 2D measurements should be blood tissue interface to blood tissue interface</p> <p>RVOT_{PLAX} $\geq 32\text{mm}$ or $\geq 19\text{mm/m}^2$ AND the presence of regional RV akinesia, dyskinesia or aneurysm is a major criterion**</p> <p>RVOT_{PLAX} $\geq 29\text{mm}$ to $< 32\text{mm}$ OR $\geq 16\text{mm/m}^2$ to $< 19\text{mm/m}^2$ AND the presence of regional RV akinesia or dyskinesia is a minor criterion**</p> |  |
| 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 | <p>Proximal RVOT (RVOT₁)</p> <p>Qualitative assessment of RV structure and function</p> <p>Regional wall motion analysis of the outflow tract of the RV (infundibulum)</p> | <p>-at end diastole*</p> <p>-measured from anterior aortic wall directly up to the RV free wall (at the level of the aortic valve)</p> <p>-the PSAX view has been shown to be more reproducible than the measurement obtained from the PLAX orientation</p> <p>RVOT₁ $\geq 36\text{mm}$ or $\geq 21\text{mm/m}^2$ in the presence of regional RV akinesia, dyskinesia or aneurysm is a major criterion**</p> |  |

| | | | | |
|---------------|----|---|--|--|
| | | | <p>RVOT1 $\geq 32\text{mm}$ to $< 36\text{mm}$ or $\geq 18\text{mm/m}^2$ to $< 21\text{mm/m}^2$ in the presence of regional RV akinesia or dyskinesia is a minor criterion**</p> | |
| PSAX PV level | 2D | <p>Distal RVOT (RVOT₂)</p> <p>Qualitative assessment of RV structure and function</p> <p>Regional wall motion analysis of the infundibulum of the RV</p> <p>PA diameter</p> | <p>-end diastole*</p> <p>-measured just proximal to PV</p> <p>There are no specific values for diagnosis of ARVC however this should be used to demonstrate dilatation.</p> <p>RVOT₂ $> 27\text{mm}$ is abnormal in other cardiac pathology*</p> <p>-end diastole</p> <p>- half way between pulmonary valve (PV) and bifurcation of main PA or 1cm distal to PV</p> <p>Enlargement of the pulmonary artery makes the diagnosis of ARVC less likely (may be indicative of conditions causing pulmonary hypertension)</p> | |
| PSAX Base | 2D | <p>Qualitative assessment of RV structure and function at basal level</p> <p>Regional wall motion analysis of inferior, lateral, anterior and septal walls of RV in PSAX at base (mitral valve) level</p> | <p>Relative size of RV to LV should be assessed</p> <p>There is disproportionate enlargement of the RV in ARVC</p> | |
| PSAX Mid | 2D | <p>Qualitative assessment of RV structure and function at papillary muscle level</p> <p>Regional wall motion analysis of inferior, lateral, anterior and septal walls of RV in PSAX at mid (papillary muscle) level</p> | <p>Relative size of RV to LV should be assessed</p> | |
| PSAX Apex | | <p>Qualitative assessment of RV structure and function at the apex</p> <p>Regional wall motion analysis of inferior, lateral and septal walls of RV in PSAX at apex level</p> | <p>Relative size of RV to LV should be assessed</p> | |

| | | | | |
|--|---------------------|---|--|--|
| 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 | |   |
| Useful additional parameters standard Apical 4CH | 2D | Basal RV:LV ratio at end diastole. 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 | 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* 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 | <ul style="list-style-type: none"> - at end diastole - ignore trabeculations and papillary muscles - use reduced depth to improve resolution and measurement accuracy <p>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.*</p> |  |
| | | IVC size and inspiratory collapse | Estimate of RA pressure to define RV end systolic pressure (see pulmonary hypertension protocol for details) |  |
| | | | |  |

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|------------|---------------------|--|---|---|
| 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., Afzal, 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 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