ANGIOGRAPHY PROCESSING AND TRANSFER

OVERVIEW

Angiography (angio) study is required, <u>if performed per standard of care</u>, for all subjects enrolled in the ARVC study. If angiography is done for diagnosis purposes at the enrolling center after consent form is signed (not previously done by outside source), they should be done using the standardized ARVC protocols in this manual. If angio was performed prior to enrollment, a copy of the latest test CD and or a copy of the test CD that was done for ARVC diagnosis purposes, should be sent along with the written report with a completed Form 4F and Form 4A. Any new angiogram done prior to a specific follow –up visit must be sent to the angio core lab as soon as it is done and the required CRFs should be completed in the subsequent yearly visit event in the ARVC electronic data entry system.

ANGIOGRAPHY PROCESSING AND PROCEDURES

If the angio is performed after subject signs the study consent form, we recommend using the angio procedures in this manual.

The angiographic features of arrhythmogenic right ventricular cardiomyopathy (ARVC) include global and/or regional function and morphological abnormalities of the right ventricle. Cardiomegaly, localized akinetic or dyskinetic bulges, outpouchings, dilatation of the infundibulum, trabecular hypertrophy and/or disarray with deep fissures are morphological findings in ARVC.

The following protocol for RV angiography is designed to illustrate these abnormalities and to perform quantitative measurements of RV volumes, ejection fraction, and regional contraction. If angiogram is performed after subject signs consent, please adhere to the following:

- Acquire good-quality RV angiogram
- Store the data on CD (DICOM format). The core lab will perform the study analyses and measurements.
- > Biplane digital catheter lab is preferred.
- Avoid ECG cables, connectors, etc in field of view.

Hemodynamic measurements (PA pressure (sys/dias/mean), RA mean pressure, and RV end diastolic pressure calibration are performed in the core lab based upon the size of the angiographic catheter used for RV angiography. Specify the type and size of catheter used: Pigtail (5Fr or larger), Berman catheter (6Fr or 7Fr).

Place the RV angiography catheter in the mid inferior RV (RAO or PA view), approximately 1 cm above the mid-inferior wall to best avoid catheter induced ectopy. Avoid contact with the RV wall or trabeculae. Confirm that the entire RV in both acquired views (i.e. the RAO-LAO acquisition, and AP/Lat acquisition) can be seen. DO NOT move the table during the acquisition. Use the slower injection rate of 12-15 ml/sec to avoid PVCs.

Projections: (preferred with biplane mode to minimize contrast needs)

- a. 30 deg RAO
- b. 60 deg LAO
- c. Postero-anterior (PA)
- d. Lateral (LAT)

Optional views: (encouraged if contrast use allows)

- a. 45 deg RAO
- b. 45 deg LAO

Image Acquisition: 30 fr/sec during a breath hold. Film long sequence to permit contrast passage to LV. Contrast load: 40-50 ml at 12-15 ml/sec.

ANGIOGRAPHY AT BASELINE

- <u>Transfer of Angiography CD and Reports for a New potential Proband:</u> The Angiography written report must be sent to the Clinical Center Reviewer for ARVC diagnosis. Once the ARVC diagnosis is verified by the Clinical Center Reviewer, the angiogram CD must be shipped to the angio core lab. It is possible that the Clinical Center Reviewer will request the angiogram images apart from the written reports to assess patient's eligibility. In this case, the Angiography CD must be sent within 5 working days from date of request to the Clinical Center Reviewer.
- 2. <u>Transfer of Angiography CD and Reports for Family Member (old/new) and **Old** Probands: Once the proband in the family is identified, or a subject (proband/family member) from the previous grant is enrolled, the Angiography CD and written report must be sent to the Angio Core lab.</u>

ANGIOGRAPHY AT FOLLOW-UP VISITS

If angiography is performed while a subject (old/new proband or old/new family member) is participating in the study, we recommend that the standardized protocol in this manual should be used. The CD and written report should be sent at the next sequential follow-up study visit following the procedure. Each sent media must be accompanied by a completed Form 4F and a completed Form 4A.

TEST MEDIA TRANSFER

The initial delivery of the image media, the forms, and the local interpretation report must be attempted by uploading the data electronically, according to instructions in transferring section of this manual. If data upload is unsuccessful, send the data to the Angio Core lab using Fed Ex.

MANUAL TRANSFER

The electronic file must be delivered to:

Dr. Julia Indik University of Arizona 1501 North Campbell Sarver Heart Center, Room 5153 Tucson, AZ 85424 USA Phone: 520-626-6262 Fax: 520-626-4333

If you have any questions, please contact Julia Indik, MD at phone number: 520-626-6262

Delivery of the report to the **Clinical Review Center** for new proband ARVC evaluation must be attempted by uploading the data electronically, according to instructions in the test transfer section. If data upload is unsuccessful, send the data via mail or fax as instructed in the test transfer section.

Table 1: Test Media Transfer

	Test Media Transferred Baseline Visit		Test Media Transferred Follow-Up Visit
Subject Category	New Proband	 Old Proband New Family Member Old Family member 	 Old Proband New Proband New Family Member Old Family member
Ship to Clinical Center Pl	 ≻Clinical Center Data Shipment Form (Form 5B) ≻Angio Form (Form 4F) ≻Written report 		
Ship to Angiography Core laboratory	 Shipping Form (Form 4A) Angio Form (Form 4F) CD Written report 	 Shipping Form (Form 4A) Angio Form (Form 4F) CD Written report 	 Shipping Form (Form 4A) Angio Form (Form 4F) CD Written report

Performance of RV Angiography Biopsy

Presentation from Thomas Wichter (Revised December 2013 – Julia Indik)



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Please respect copyright of all RV angiogram images and movies displayed in this demo presentation which was designed for training purposes within the NIH-funded "Multidisciplinary Study of ARVC".

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(Thomas Wichter, MD, FESC).

ANGIOGRAPHY PROTOCOL FOR ARVC

The diagnosis of arrhythmogenic right ventricular dysplasia (ARVC) is made on angiography by showing decreased global and/or regional function and illustrating morphological abnormalities of the right ventricle. Cardiomegaly, localized akinetic or dyskinetic bulges, outpouchings, dilatation of the infundibulum, trabecular hypertrophy and/or disarray with deep fissures are morphological findings in ARVC.

> Multidisciplinary Study of ARVC Angiography Core Lab

ANGIOGRAPHY PROTOCOL FOR ARVC

The following protocol for RV angiography is designed to illustrate these abnormalities and to perform quantitative measurements of RV volumes, ejection fraction, and regional contraction. It will be used uniformly within the multidisciplinary study of ARVC. Please make sure to acquire good-quality RV angiograms stored on CD. We will take care of the study analyses and measurements.













RV-angio in 30° RAO view: Trabecular hypertrophy ant-sept, polycyclic contours infero-apical



RV-angio in 30° RAO view: RV enlarged, dilated RVO, akinetic inferior wall, tricuspid regurgitation















RV BIOPSY PROTOCOL FOR ARVC

The histological diagnosis of arrhythmogenic right ventricular dysplasia (ARVC) is made from histological slides obtained from right ventricular endomyocardial biopsy samples. Typical histological features of ARVC include myocardial atrophy with replacement by fibrous and/or fatty tissue interspersed with surviving strands of hypertrophied myocytes. These changes affect primarily the RV free wall whereas the interventricular septum is usually spared. Multidisciplinary Study of ARVC

Angiography Core Lab

RV BIOPSY PROTOCOL FOR ARVC

Because the septum is usually spared and because tissue abnormalities may be localized, there is significant sampling error if standard septal sampling is performed. Predilection areas in the subtricuspid (basal inferior), apical and outflow tract areas of the right ventricular free wall. Therefore, biopsies should be sampled at the myocardial border of the RV free wall and the septum at the areas of predilection or documented contraction abnormalities.

The following protocol for RV biopsy is designed to provide a description of RV biopsy technique in ARVC for the use within the multidisciplinary study of ARVC.



Protocol for RV Biopsy (2)

Use bioptome with medium sized jaws

Create 70-90° smoothe curve at distal tip (5-7 cm) of bioptome

Take 5 samples of sufficient size (see pathology Core-Lab)

Record RV and RA pressures after sampling is finished







70-90° smoothe distal curve Bioptome in long sheath

Bioptome jaws (med. size)



