

## Invitation to Participate

An invitation to participate in a multicenter prospective coordinated database registry on anomalous origins of a coronary artery from the opposite sinus of valsalva (ACAOS), with interarterial course or an intramural aortic course.

## ACAOS MULTICENTER STUDY

### Summary

In order to better subclassify ACAOS carriers and their medical histories, along with the results of interventions, a database has been established with the following characteristics:

- a) openness to any physician in the world interested in collaborating
- b) defined entry characteristics
- c) defined protocols
- d) total disclosure (to database) of events and scheduled follow-up methods
- e) privacy protection
- f) commitment to research and education

### Study Objectives

Of all congenital coronary anomalies, only a few continue to present confusing evidence of their clinical relevance and carry a potential for catastrophic (or at least, important) events; they are mainly comprehended in the group of “**ACAOS**,” **A**nomalous origin of a **C**oronary **A**rtery from the **O**pposite **S**inus of **V**alsalva, with intramural aortic proximal course, and/or interarterial course.

We are attempting to gather adequate information in a large group of carriers, in order to clarify natural prognosis and results of interventions. For “Guidelines for Acceptable Case Studies”, please see page 6 of this document.

### *Who should be interested in collaborating?*

Cardiologists or cardiovascular surgeons who come to identify, by any means, carriers of ACAOS, and who are willing to participate in this multicenter research endeavour to promote a worthwhile research project.

### *What is the benefit to the outside collaborating investigator?*

The investigators will be able to:

- share in the experience of involved colleagues.
- influence the investigation itself (by submitting observations/advice).
- contribute to the publication of the final report (every participant will be recognized in a “list of investigators,” and those who supply significant support will also be recognized in the “list of authors”).
- as a participating physician, obtain (free) expert consultation on individual cases.

*How does it work?*

A physician who has identified at least 1 case of ACAOS (or many cases) initially submits an application to participate and is invited to register using the “CAAC Society Registration Form For Participating Physician’ (see page 8). After enrolling in the study, the participating physician should follow the disciplined practice of including all patients who are carriers of ACAOS in his/her professional practice, not just the most interesting or complicated cases, or the ones that were intervened. Each case/patient should be entered individually by filling out the appropriate “Individual Patient Database Entry Form”(see page 3). Each patient should be followed up, as outlined in the “Individual Patient Database Entry Form” follow-up protocols.

*Participating physician protection:*

Each investigator is assured that:

- a) the privacy of each patient is strictly protected (no name will ever be mentioned). Each patient is identified in the database by initials and birth date only (e.g. “P.A. 01.09.41”).
- b) no case will be entered in a published report without acknowledging the responsible physician’s contribution.

**Individual Patient Database Entry Form**  
(will require approximately 10 minutes)

The participating physician is invited to send the following information:  
(include as much as available, but a minimum is required to qualify for entry)

**Print and mail this completed form to:**

Paolo Angelini, MD  
CAAC  
P. O. Box 20206  
Houston, TX 77225  
Phone 713-790-9401  
Fax 713-790-0353

e-mail: [leachman@leachmancardiology.com?subject=CAAC website](mailto:leachman@leachmancardiology.com?subject=CAAC%20website)

a) **Patient name (initials only):** \_\_\_\_\_

b) **Date of birth:** \_\_\_\_\_

c) **Sex:** \_\_\_\_\_

d) **History of athletic activities** (Type? When? Briefly describe.):

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

e) **Date of initial entry into database** (today): \_\_\_\_\_

f) **Coronary anomaly diagnosis:** \_\_\_\_\_

Associated conditions: \_\_\_\_\_

\_\_\_\_\_

Confirmation at CCAC: yes \_\_\_\_\_ no \_\_\_\_\_

(if not confirmed, new diagnosis: \_\_\_\_\_)

g) **Presenting symptoms** (circumstances presented to first physician who identified condition of ACAOS):

	<b>yes</b>	<b>no</b>
• angina:		
i. typical (effort-related)	_____	_____
ii. typical for location/radiation	_____	_____
iii. atypical (non-effort-related)	_____	_____

- iv. atypical for location/radiation \_\_\_\_\_
- dyspnea: functional class \_\_\_\_\_
- palpitations \_\_\_\_\_
- syncope (times) \_\_\_\_\_
- cardiac arrest:
  - i. aborted \_\_\_\_\_
  - ii. terminal \_\_\_\_\_
  - iii. during effort (mild, moderate, extreme) \_\_\_\_\_
  - iv. after effort \_\_\_\_\_
  - v. non-effort-related \_\_\_\_\_

**h) Diagnostic work up:**

1. EKG: normal/abnormal (send copy if positive)
2. ECHO: TTE, TEE (send report) Double product: \_\_\_\_\_
3. Stress Tests:
  - Treadmill +, -, +/- Double product: \_\_\_\_\_
  - Cycloergometer +, -, +/- Double product: \_\_\_\_\_
  - Nuclear imaging + scarring (where?): \_\_\_\_\_
  - + ischemia (where?): \_\_\_\_\_
4. Cardiac MRI: diagnostic findings (send report)
5. Ultrafast CAT scan: diagnostic findings (send report)
6. Coronary angiography: send report + CD-R/by e-mail)
7. IVUS: (send report / CD-R) or not done

**i) Therapy:**

- None (no medicines, no physical limitations) \_\_\_\_\_
- Physical limitations imposed: I(none) II(athletic activities) III(major exertion):  
\_\_\_\_\_
- Medications:
  1. beta-blockers: \_\_\_\_\_
  2. anti-hypertensive: \_\_\_\_\_
  3. calcium antagonists: \_\_\_\_\_
  4. nitrates: \_\_\_\_\_

**j) Interventions:**

1. Stent angioplasty: \_\_\_\_\_  
\_\_\_\_\_
  - a) at intramural anomalous course
  - b) at atherosclerotic disease, remote from intramural segment

2. Surgery: \_\_\_\_\_  
\_\_\_\_\_

- a) bypass: IMA to anomalous artery or vein graft to anomalous artery
- b) relocation of ostial cuff
- c) ostioplasty (creation of new ostium, without relocation)
- d) unroofing (from ostium)
- e) other (describe): \_\_\_\_\_

**k) Follow-up since interventions** (suggested: once a year, unless new symptomatic events):

- Symptoms (copy from list above): \_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_
- Changes: \_\_\_\_\_
  - i. Improved
  - ii. Same
  - iii. Worse
  - iv. Totally fine (for cardiac symptoms)
- New procedures (date, type, see above): \_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_
- New medicines (date, type, see above): \_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_
- Death:
  - i. sudden cardiac (date) \_\_\_\_\_
  - ii. non-cardiac (date) \_\_\_\_\_
  - iii. ACAOS-related (date) \_\_\_\_\_
  - iv. Others \_\_\_\_\_
- Other Comments:

**Guidelines for Acceptable Case Studies**  
**Classification of Pertinent Types of Anomalous Origin of a Coronary Artery**  
**(FOR MULTICENTER “ACAOS DATABASE REGISTRY”)**

At this time, for the aims of the ACAOS project, we are only interested in the following anomalies/characteristics:

- A. Ectopic coronary ostium from:
- a. Ascending aorta (above sinutubular junction)
  - b. Coronary sinuses - Proper sinus, abnormal location (high, low, commissural)
    - Non-coronary sinus (NCS)
    - Opposite coronary sinus

Subtypes:

1. RCA from Left Sinus or LCA
2. RCA from NCS
3. LCA from R sinus or RCA
4. LCA from NCS
5. LAD from R sinus or RCA
6. CX from R sinus or RCA
7. CX form NCS

Note: At this time we will not collect other anomalies, like cases of muscular bridges, anomalous origin from the pulmonary artery, fistulae, etc.

- B. When applicable, the above types should be further subclassified by “Proximal course of the ectopic vessel”:
- a. Anterior (pre-pulmonic)
  - b. Intraseptal (ventricular septum)
  - c. Interarterial (between aorta/pulmonary artery sinuses)
  - d. Retro-aortic

Note: We will not collect cases of posterior course (CA from RCA, distal or RCA from CX, distal) since this case is quite frequent and not clearly associated with clinical repercussions.

If IVUS (intravascular ultrasound study) was performed, and intussusception of ectopic artery at aortic wall was found, then enter the following parameters:

- Intussusception: present or absent
- Stenosis of such segment: area narrowing (% of distal lumen, reference vessel) \_\_\_\_\_
- Hypoplasia of such segment (area of external elastic membrane as % of distal reference vessel) \_\_\_\_\_
- Intimal thickening (plaque area at such segment): yes or no \_\_\_\_\_
- Intimal thickening at distal segment (adjacent to intussuscepted): mild, moderate, or severe \_\_\_\_\_

## MANUSCRIPT PREPARATION

In case a physician would like to submit a case report for publication in the Texas Heart Journal, under the “Coronary Anomalies” section, we suggest they follow the Routine Guidelines for Authors (See Journal website at [www.texasheartinstitute.org](http://www.texasheartinstitute.org)\*), and observe the following recommendations:

- a. Case reports should be short and mainly concerned with individual case peculiarities (clinical presentation, imaging, intervention, etc.).
- b. Introduction and discussion should not repeat generalities (incidence of coronary anomalies, embriology, etc.), but refer to existing reviews (chapter of book/books/ fundamental review articles) on coronary anomalies.
- c. Specific mention should be made in order to document the:
  - i. presence of typical and atypical chest pain.
  - ii. presence of dyspnea, syncopal equivalent, sudden death.
  - iii. objective evidence of coronary anomaly-related reversible ischemia (echo stress or nuclear scintigraphy) or scarring.
  - iv. follow-up under specific treatment (including recommendation of sport activities to curtail).

**Print and mail completed manuscript to:**

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e-mail: [leachman@leachmancardiology.com?subject=CAAC website](mailto:leachman@leachmancardiology.com?subject=CAAC website)

CAAC SOCIETY REGISTRATION FORM  
FOR PARTICIPATING PHYSICIAN

Name: (last) \_\_\_\_\_, (first) \_\_\_\_\_

Mailing Address: (street) \_\_\_\_\_

(city) \_\_\_\_\_

(country) \_\_\_\_\_

E-mail: \_\_\_\_\_

Telephone: \_\_\_\_\_

Fax: \_\_\_\_\_

Specialty: \_\_\_\_\_  
(cardiologist, CV surgeon, others)

Institution where practicing: \_\_\_\_\_

(location) \_\_\_\_\_

MD, DO: \_\_\_\_\_  
(graduation year and Institution )

Academic institution to which the physician is associated and title:

\_\_\_\_\_  
(instructor, professor, etc.)

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