Remarks on Physiopathogenesis

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American “Legion of Honor” 1/5 2005 (#3)
Selection made by 34,000 researchers around the World
after 3 decades of investigation, all categories included

British “Order of Distinction” 1/400

Orlando 11/07
I have been able to identify ARVD because of my expertise in Clinical Electrophysiology namely the surgical treatment of the Chronic Forms of Ventricular Tachycardia

To understand the Electrogenesis of late potentials I was led to study histology of RV from Surgical Samples

For their proper interpretation I trained during a 10 Year period with a Heart Pathologist (Dr Fabrice Fontaliran, Paris)

Fontaine et al. in Zipes Book 2004
**Arrhythmogenic Right Ventricular Dysplasia**

**Typical Clinical History**

- Lets start by reporting a typical well documented patient
- Diagnosis suspected after recording of Exercise induced Ventricular Tachycardia with LBBB pattern
- This patient will go from normalcy to Heart Transplant in 30 months

*Fontaine Unpublished 2007*
Arrhythmogenic Right Ventricular Dysplasia

A typical example of ARVD > Myocarditis > Heart Tx


Male 28 No personal history, Mother presence of PVCs

May 1998 : Palpitations during soccer
September 1998 : Near syncopal VT documented
    - Normal examination
    - Echo Dilated RA
    - Moderate RV dilatation
    - Late potentials +++
    - MRI Dilated RV + Hypersignal
    - Radio-Nuclide : LVEF 65%
    - RV infundibular Aneurysm
    - Increased thickness of trabeculations
Arrhythmogenic Right Ventricular Dysplasia

A typical example of ARVD Myocarditis > Heart Tx

End September 1998:
- Chest pain
- ST segment elevation in II, III, AVF
- Troponin 1C : 120+++ 
- Para Influenza III : borderline
- Rickettsia Conorii : borderline
- Diagnosis of Pericardo-myocarditis

October 1998: Troponin IC : 8

December 1998:
- Para Influenza III : Positive
- Radionuclide : LVEF 57%
- Increase of RV size
Arrhythmogenic Right Ventricular Dysplasia

A typical example of ARVD Myocarditis > Heart Tx

April 1999:
- Severe episode of VT with syncope despite Sotalol
- ICD implantation
- Troponin 1C : 43

February 2000: Troponin : 10

December 2000:
- Radionuclide LVEF 45%
- Troponin : 58

February 2001: Troponin : 41

March 2001:
- Radionuclide LVEF 25%
- Echo : Thrombus intra LV
- Major signs of liver dysfunction
Arrhythmogenic Right Ventricular Dysplasia

A typical example of ARVD Myocarditis > Heart Tx

March 2001 : Heart Transplant
- Histology :
  - Typical ARVD Fibrofatty replacement in RV
  - Acute signs of inflammation Lymphocytes in RV :
    - Acute signs of multi focal chronic-active myocarditis with lymphocytes in LV. No significant fat in LV

After Transplantation :
- Persistence of signs of liver failure during one month

- Obvious signs of clinical Improvement
Arrhythmogenic Right Ventricular Dysplasia

A typical example of ARVD > Myocarditis > Heart Tx

Conclusion:
- Typical clinical case of ARVD with severe ventricular arrhythmias
- Acute signs of clinical inflammation
- LVEF Drop from 57 to 25% in 27 months!

Histologic Material shows:
- Fibrofatty replacement in RV > ARVD
- Acute signs of multi focal chronic-active myocarditis with lymphocytes in LV
  explaining biventricular failure and successful Tx indication
Arrhythmogenic Right Ventricular Dysplasia

Effect of Myocarditis on LVEF over Time

Fontaine et al Circulation (letter) 1988
Arrhythmogenic Right Ventricular Dysplasia

LVEF in ARVD

Fontaine et al Heart & Vessels 1988
Arrhythmogenic Right Ventricular Dysplasia

Recent series of 22 ascertained ARVDs

Fontaine et al in Marcus Book 2007
Arrhythmogenic Right Ventricular Dysplasia

Acute Myocarditis followed by Replacement Fibrosis

- Female 45
- Clinical ARVD
- No Ventricular Tachycardia some PVCs
- Progressive Heart Failure
- Heart Tx

Fontaine et al Arch Mal Coeur 1987
Arrhythmogenic Right Ventricular Dysplasia

Right Ventricle
Arrhythmogenic Right Ventricular Dysplasia

Transmural LV Replacement Fibrosis
Arrhythmogenic Right Ventricular Dysplasia

Same Patient LV Acute signs of Myocarditis
Arrhythmogenic Right Ventricular Dysplasia

Myocarditis can be due to Bacteria

- Male 22
- Sudden Death during competitive Soccer
- Histology:
  - RV Typical ARVD
  - LV on the next picture: Macrophages Polymorphonuclears

Fontaine Unpublished 2007
Arrhythmogenic Right Ventricular Dysplasia

Transmural LV Abcess
Arrhythmogenic Right Ventricular Dysplasia

Transmural LV Abscess
Arrhythmogenic Right Ventricular Dysplasia

Differential Diagnosis
Subendocardial Myocarditis

- Male 26
- Sudden Death during recreational soccer
- Clinical Diagnosis of ARVD
- VT of RV origin
- LV multifocal zones of acute and healed myocarditis

Fontaine Unpublished 2007
Arrhythmogenic Right Ventricular Dysplasia

Differential Diagnosis
Subendocardial Myoccarditis
Arrhythmogenic Right Ventricular Dysplasia

Biventricular Dysplasia

- Girl 16
- Heart Tx for progressive Heart Failure
- No Ventricular Tachycardia
- Younger Brother also transplanted for the same reason

Fontaine Unpublished Data 2007
Arrhythmogenic Right Ventricular Dysplasia

Clinical Form
Biventricular Dysplasia

Right Ventricle

Left Ventricle

Transformation Adipocytaire
Biventriculaire
(Face Ant VD-VG)

Transformation Adipocytaire
(Coupe VG)
LV involvement in ARVD can be due to:

1. Myocarditis LV Involvement is Prognosis determinant

2. Biventricular Dysplasia with Major loss of LV Myocytes

Combination of genetic and environmental factor is common in human pathology

Original VT could have been triggered by inflammation

Fontaine 2007
What is first? There is the possibility of the opposite mechanism abnormal myocardium attracting viruses

GF answer: I have two cases to answer this question!

1. In a 27 weeks old fetus with ascertained ARVD I found fat and fibrosis but no sign of inflammation

2. I have the case of a young girl who died by accident I saw fibrosis and fat but no sign of inflammation

There is a probable bias in the myocardium of sudden death of ARVD Cases published by Domenico in the JACC because myocarditis can be by itself the trigger of sudden death...
Biventricular failure is not always the final presentation.

Huge RV dilatation at the end stage of the disease can be observed with preserved LVEF (Case Rav).

We have to take into account LV Compression by dilated RV which reduces indirectly LVEF.

Fontaine et al. Arch Mal Coeur 2006
Autopsy proven case of Hyperacute Myocarditis superimposed on typical mediomural fibrofatty replacement leading to Fulminant heart failure and death in 16 hours in a 14 months old baby.

Two other cases in the siblings at age 13 and 16 months old (one autopsied>Myocarditis+ARVD).
THANK YOU!

For more Calligraphies and ARVD discussion visit the site

http://guyfontaine.com

ARVD Corner (Drs)
ARVD News (Patients)
ARVD Forum (Patients)

Calligraphic Painting
Oil & Acrylic
on Wood
120x30

by Guy Fontaine 1964
# 126/161

“The Spring” (La Source) after Brancusi & Ingres
Louvre Museum, Paris

La Salpêtrière

ARVD LOGO
after WHF LOGO

Department LOGO

Calligraphic Painting
Oil & Acrylic
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by Guy Fontaine 1964
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