



# ARVCs in Orlando 07



## *Remarks on Physiopathogenesis*

**Guy Fontaine** MD PhD HDR FAHA FACC FESC FHRS

**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

# *Arrhythmogenic Right Ventricular Dysplasia*

---

- ◆ 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

## 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

# *Arrhythmogenic Right Ventricular Dysplasia*

---

## A typical example of ARVD > Myocarditis > Heart Tx

Medical Thesis: Fabrice Rolland : Toulouse, 2002

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 !

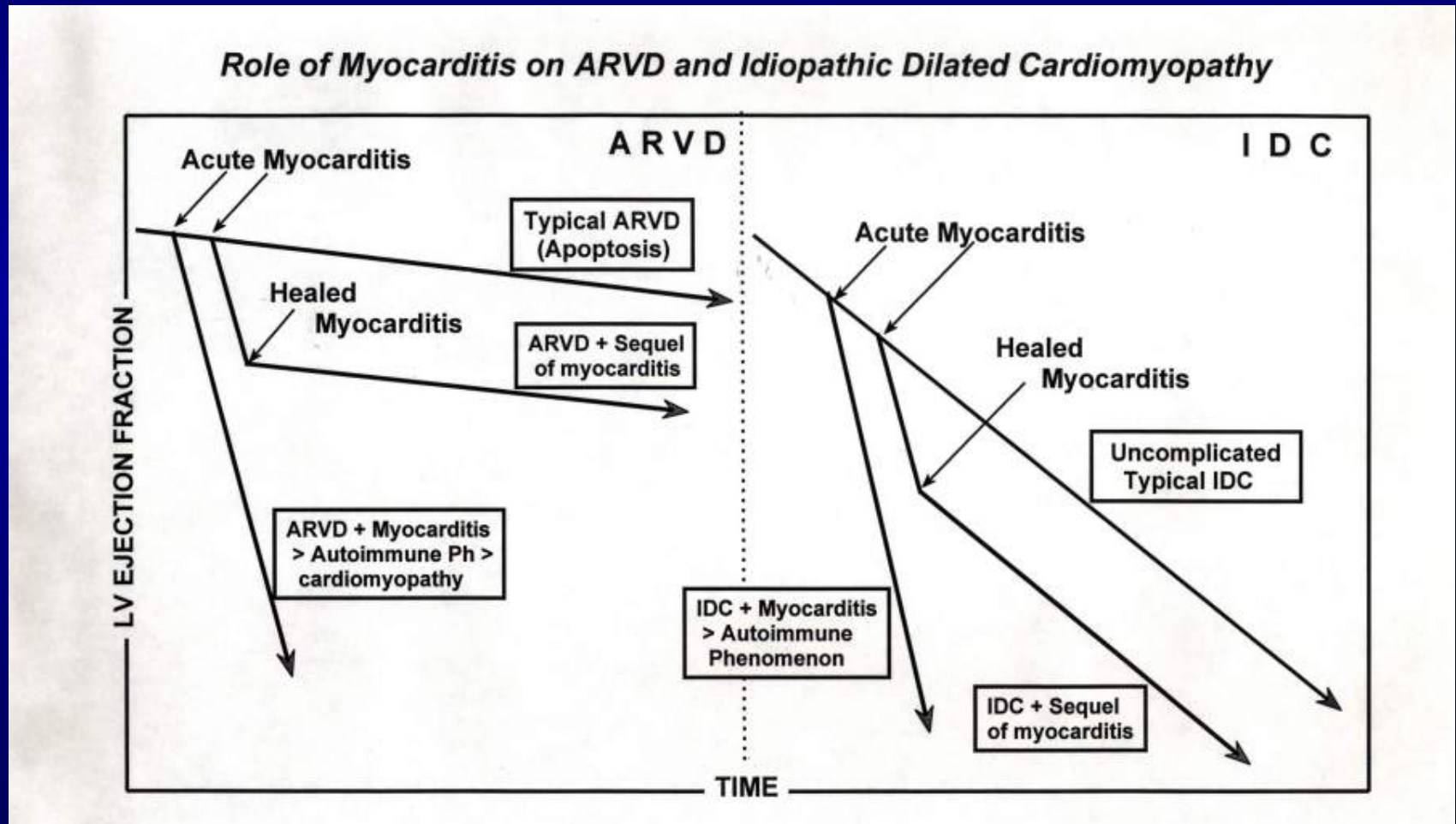
### Histologic Material shows :

- Fibrofatty replacement in RV > ARVD
  - Acute signs of multi focal chronic-active myocarditis with lymphocytes in LV
- explaining biventricular failure and successful Tx indication



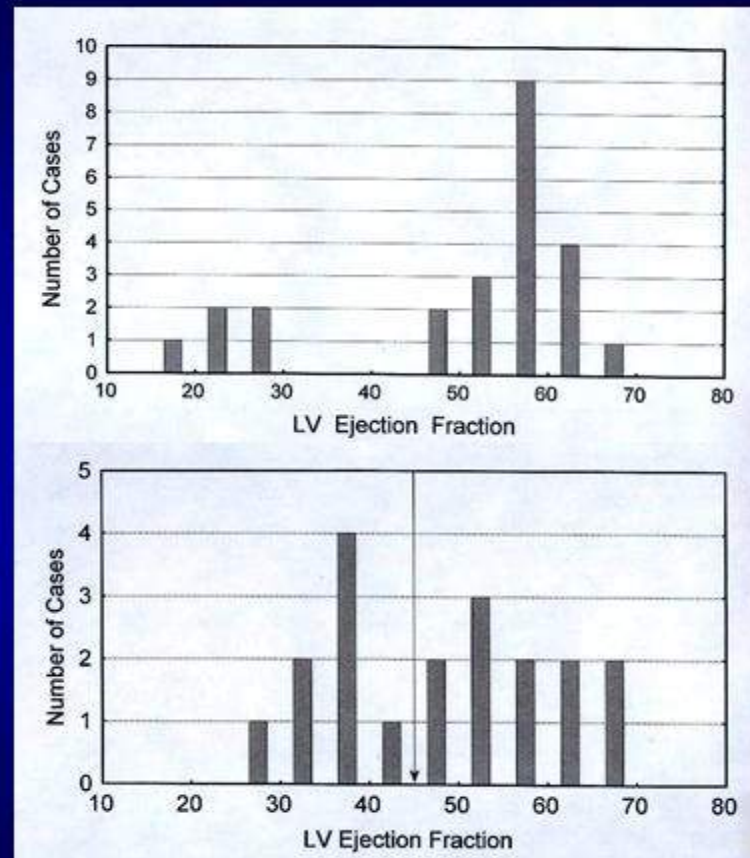
# Arrhythmogenic Right Ventricular Dysplasia

## Effect of Myocarditis on LVEF over Time



# Arrhythmogenic Right Ventricular Dysplasia

## LVEF in ARVD

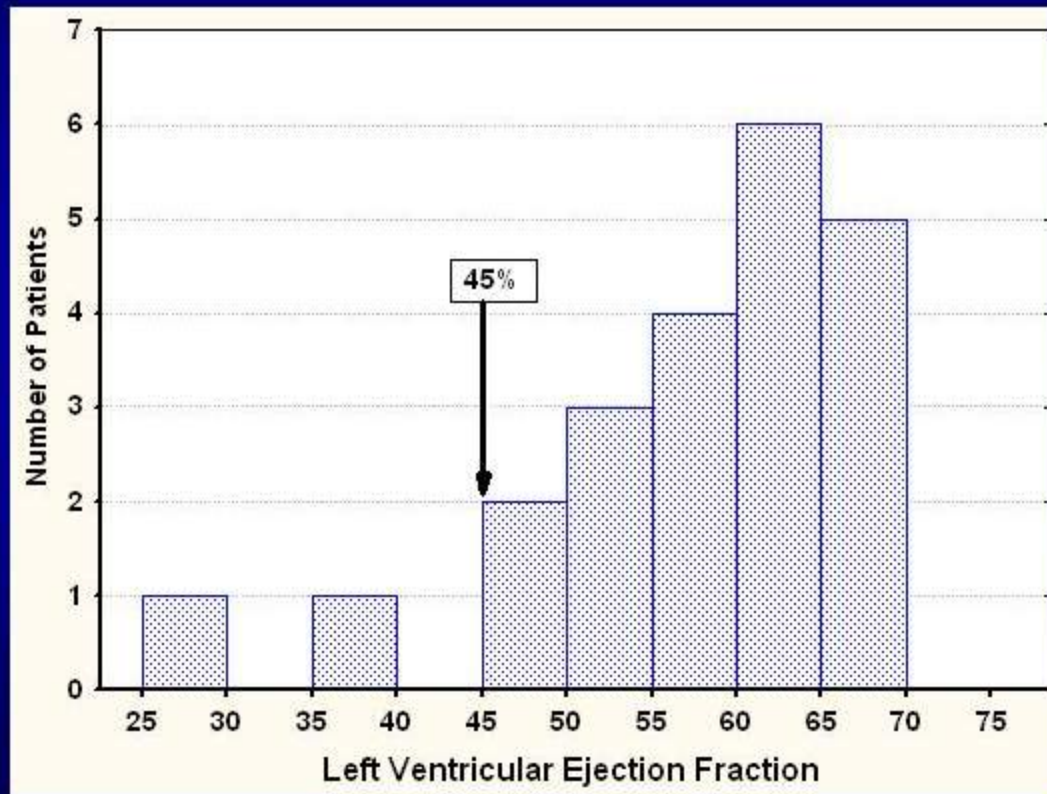


Personal  
Cohort

All  
Literature

# *Arrhythmogenic Right Ventricular Dysplasia*

Recent series of 22 ascertained ARVDs



---

*Arrhythmogenic Right Ventricular Dysplasia*

Acute Myocarditis followed by  
Replacement Fibrosis

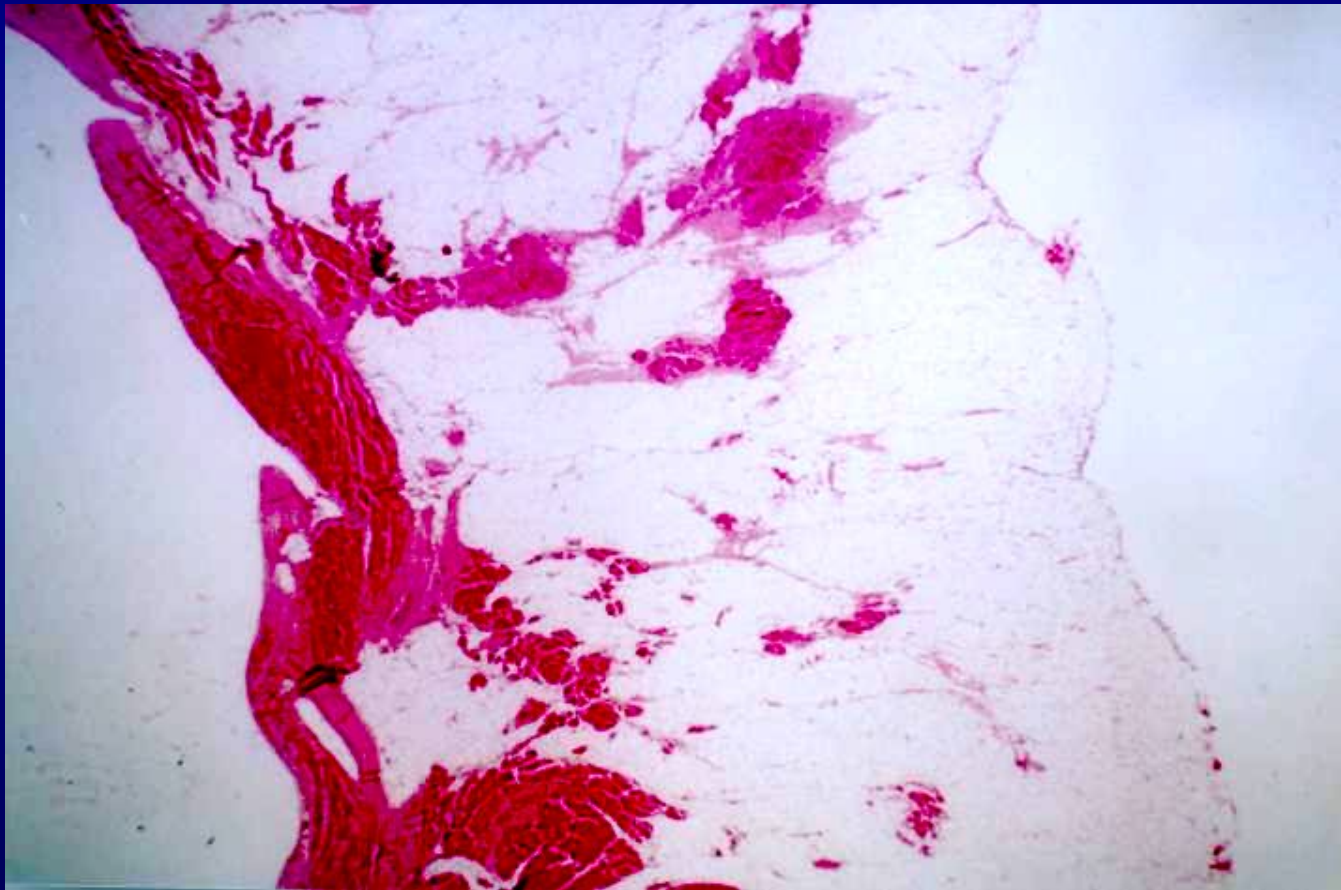
- ◆ Female 45
- ◆ Clinical ARVD
- ◆ No Ventricular Tachycardia some PVCs
- ◆ Progressive Heart Failure
- ◆ Heart Tx

Fontaine et al Arch Mal Coeur 1987

# *Arrhythmogenic Right Ventricular Dysplasia*

---

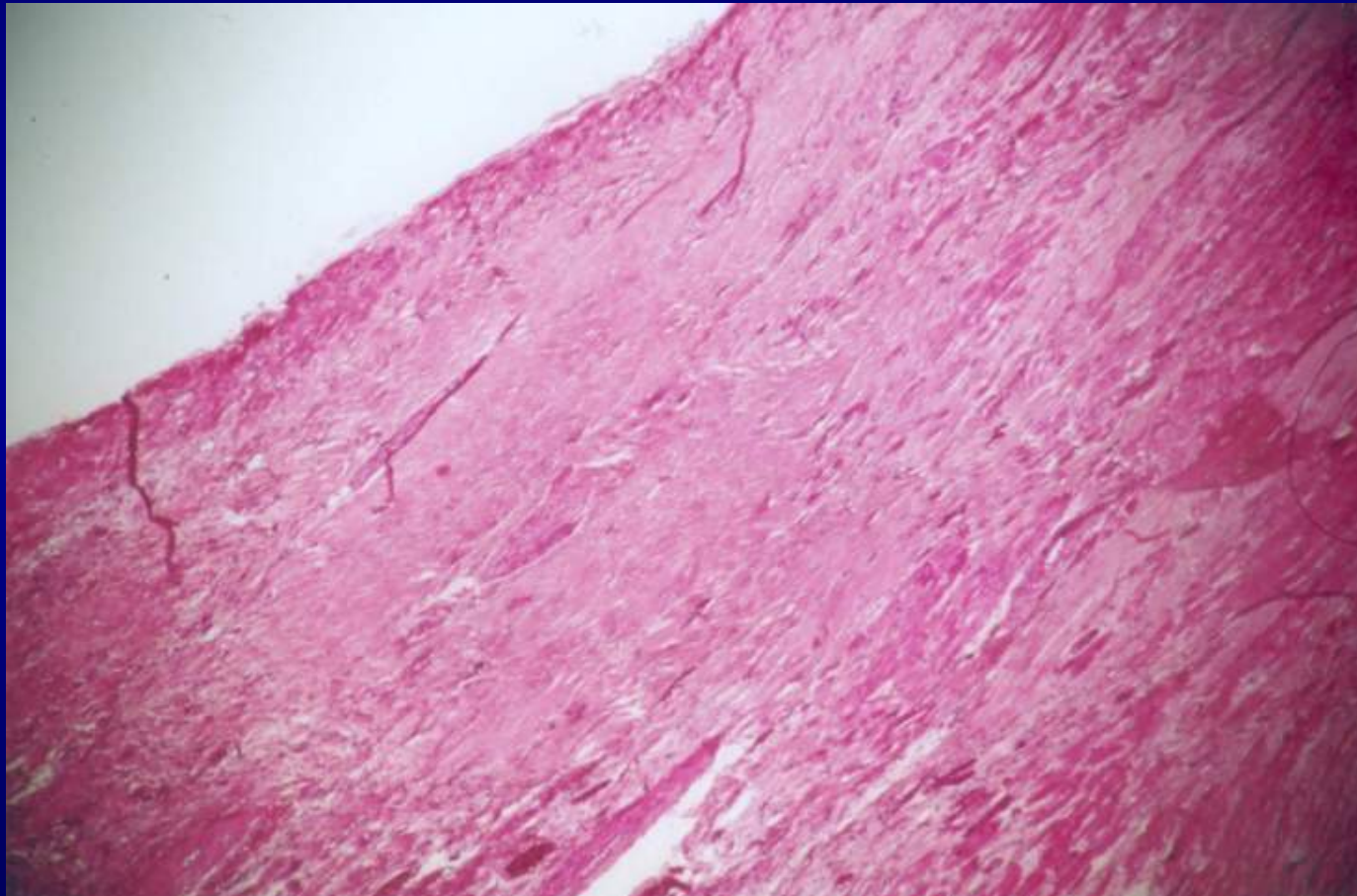
## Right Ventricle



# *Arrhythmogenic Right Ventricular Dysplasia*

---

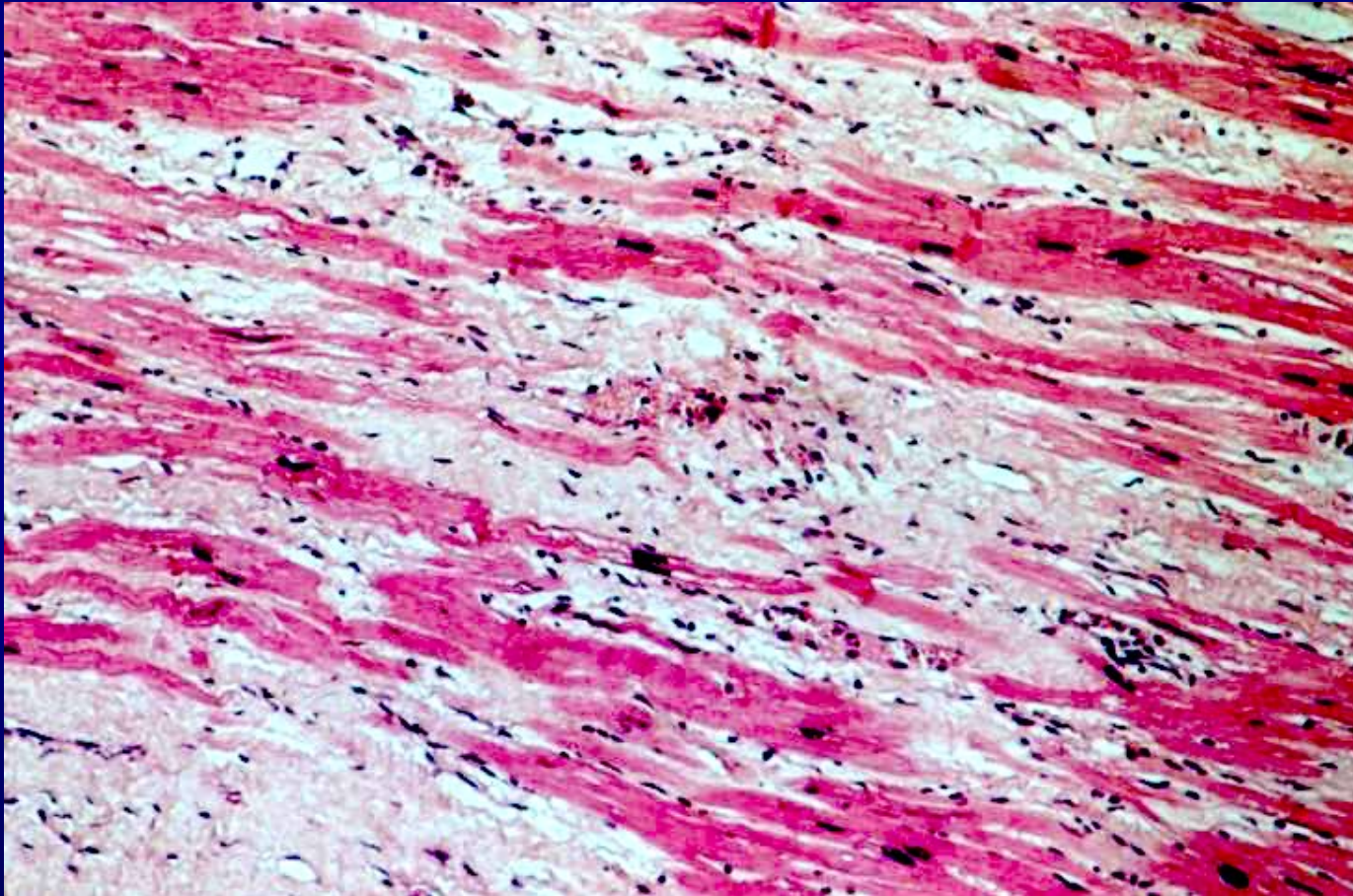
## Transmural LV Replacement Fibrosis



# *Arrhythmogenic Right Ventricular Dysplasia*

---

Same Patient LV Acute signs of Myocarditis



Myocarditis can be due to Bacteria

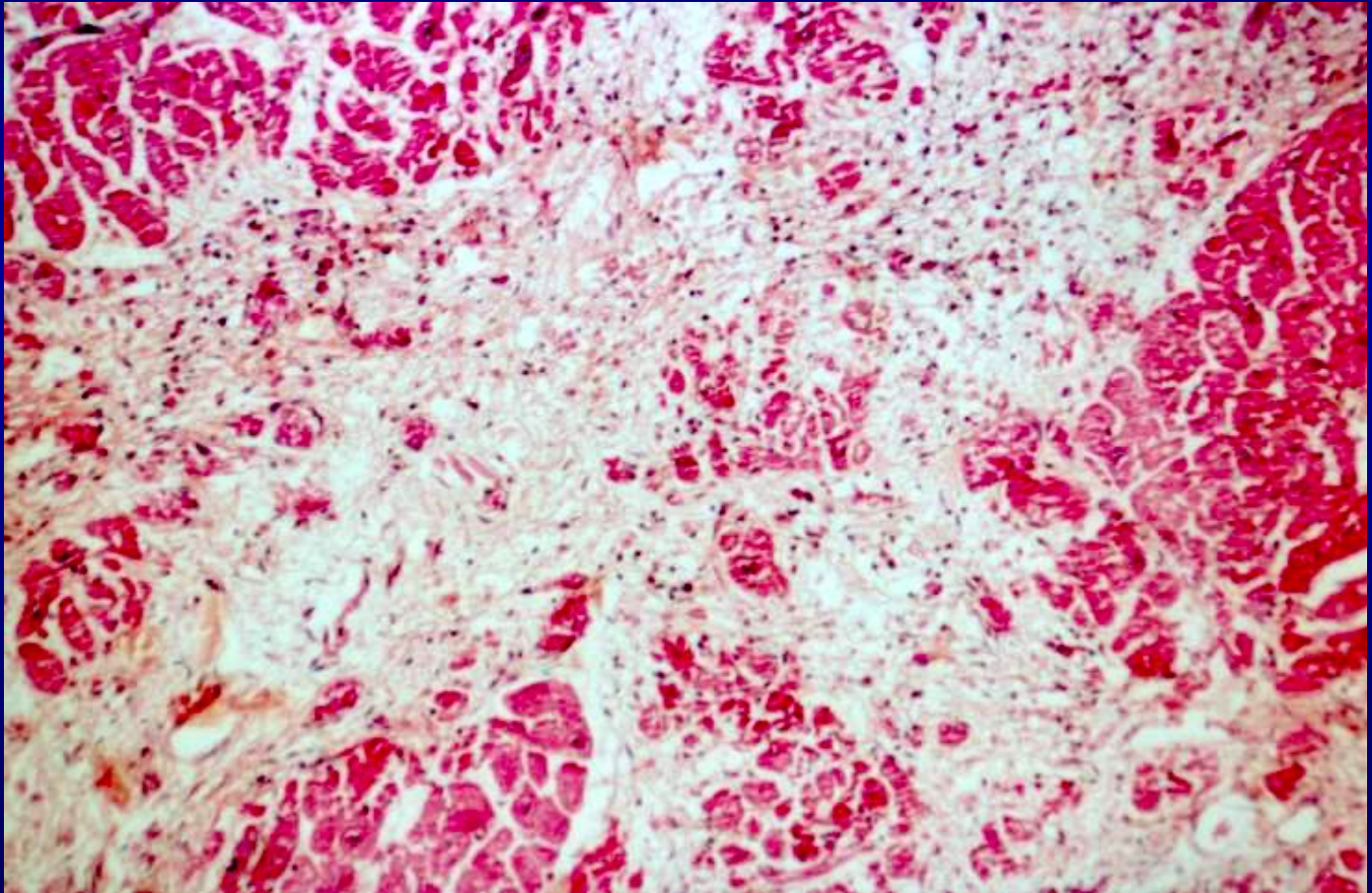
- ◆ Male 22
- ◆ Sudden Death during competitive Soccer
- ◆ Histology :
- ◆ RV Typical ARVD
- ◆ LV on the next picture : Macrophages  
Polymorphonuclears



*Arrhythmogenic Right Ventricular Dysplasia*

---

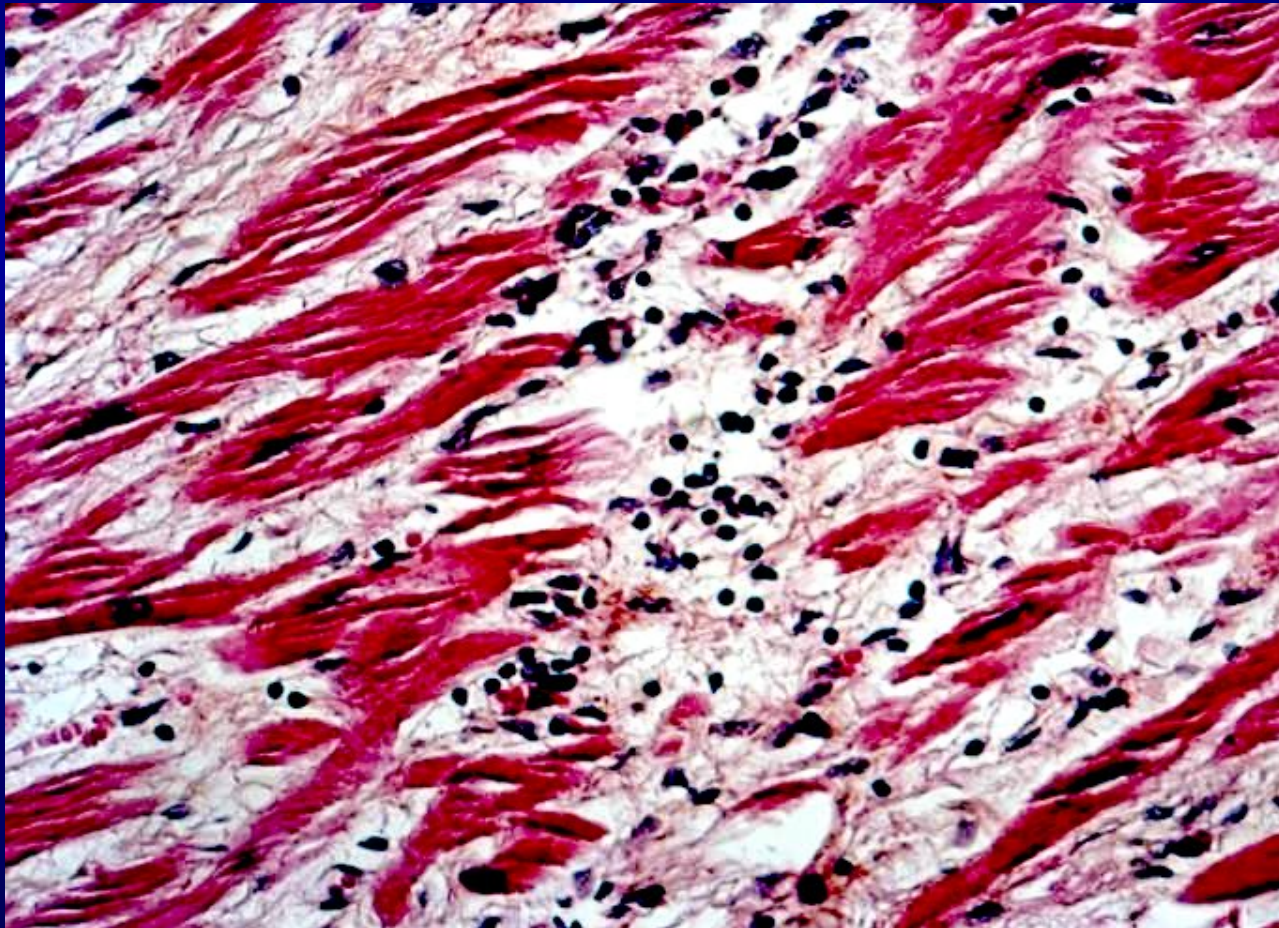
**Transmural LV Abcess**



*Arrhythmogenic Right Ventricular Dysplasia*

---

**Transmural LV Abscess**



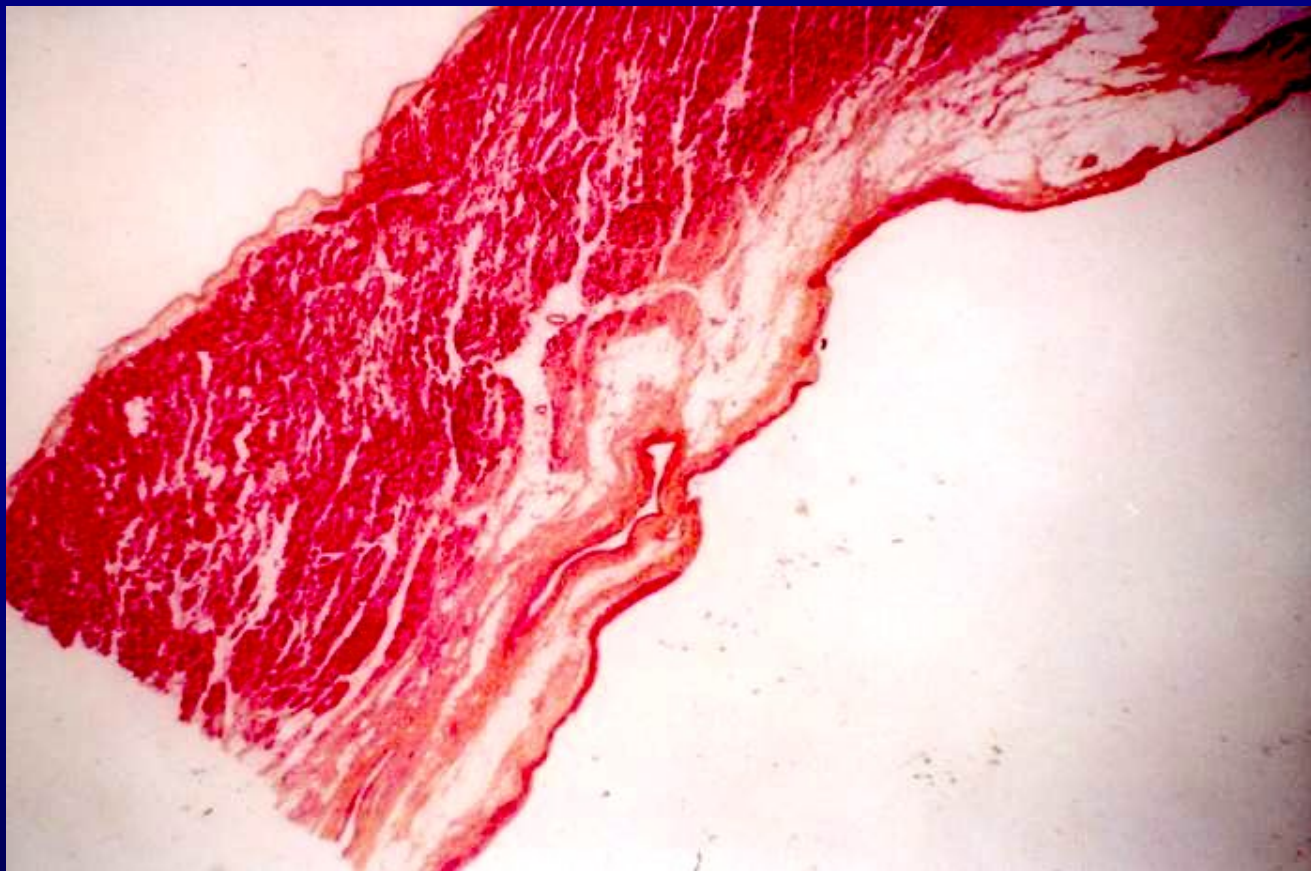
Differential Diagnosis  
Subendocardial Myocarditis

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

# *Arrhythmogenic Right Ventricular Dysplasia*

---

## Differential Diagnosis Subendocardial Myocarditis



## Biventricular Dysplasia

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

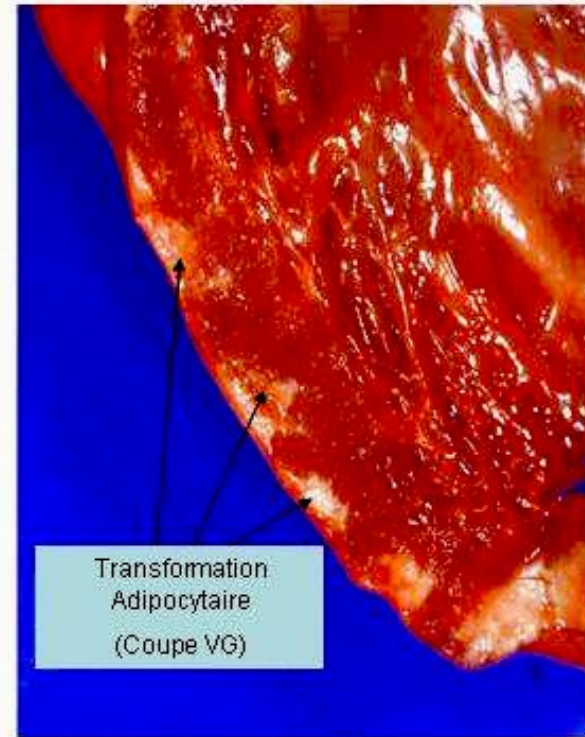
# *Arrhythmogenic Right Ventricular Dysplasia*

## Clinical Form Biventricular Dysplasia

**Right Ventricle**



**Left Ventricle**



## Conclusion

- ◆ 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

# *Arrhythmogenic Right Ventricular Dysplasia*

---

## Discussion Cristina Basso (Padoua)

- ◆ 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...



## Additional Comment 1

- ◆ 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

Additional Comment 2  
The opposite

- ◆ 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).



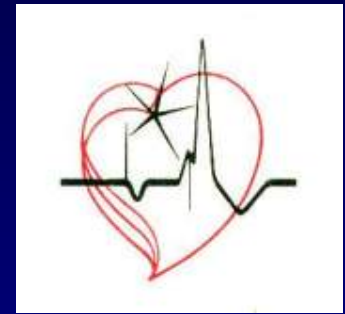
*ARVD LOGO  
after WHF LOGO*

**THANK  
YOU !**

For more Calligraphies  
and ARVD discussion  
visit the site

<http://guyfontaine.com>

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



*Department LOGO  
La Salpêtrière*

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

*Calligraphic Painting  
Oil & Acrylic  
on Wood  
120x30*

*by Guy Fontaine 1964  
# 126/161*