

# 6 Χρόνια CHALLENGE: Πού βρισκόμαστε το 2018;



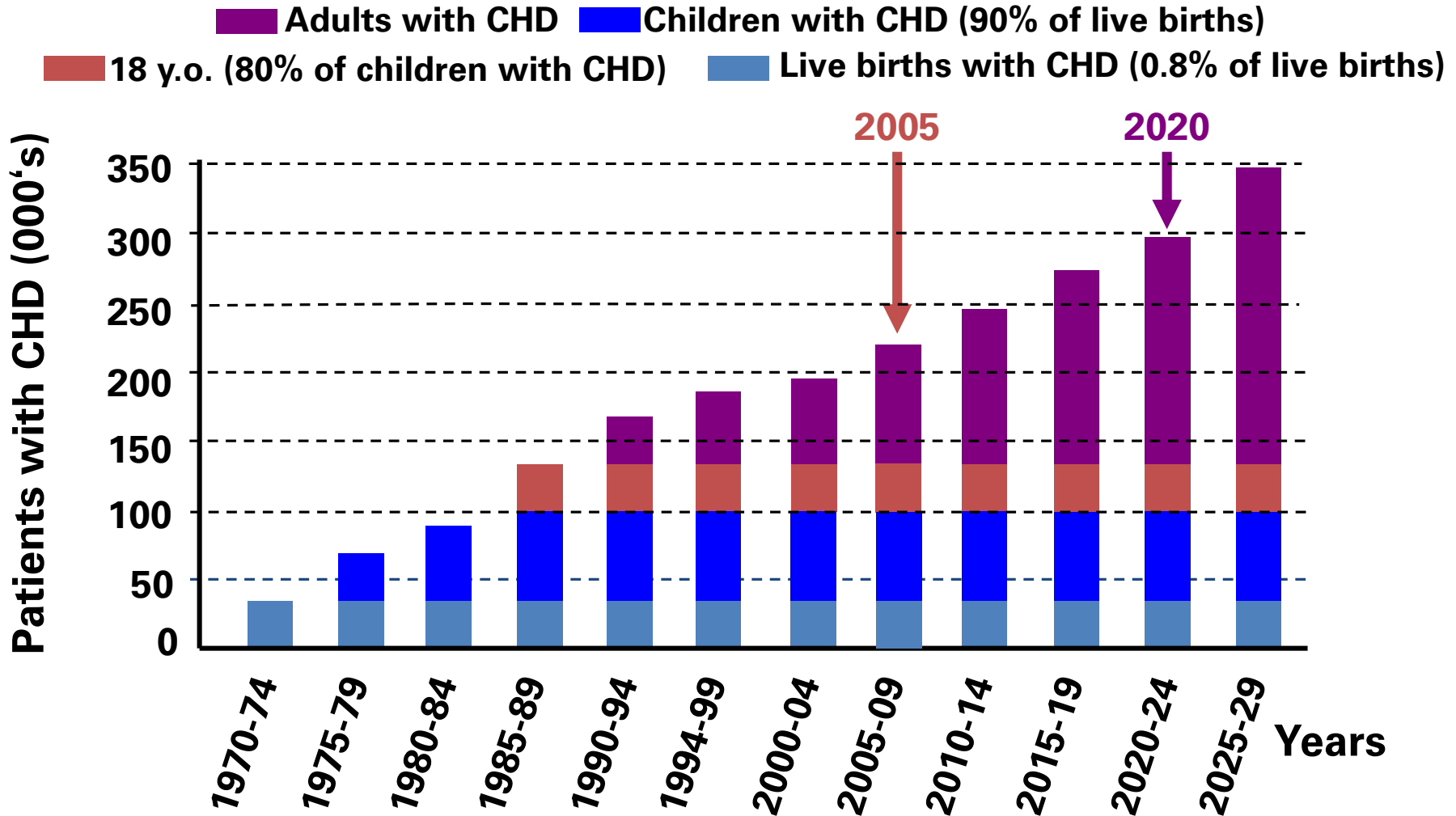
Γεώργιος Γιαννακούλας

Επίκουρος Καθηγητής Καρδιολογίας ΑΠΘ

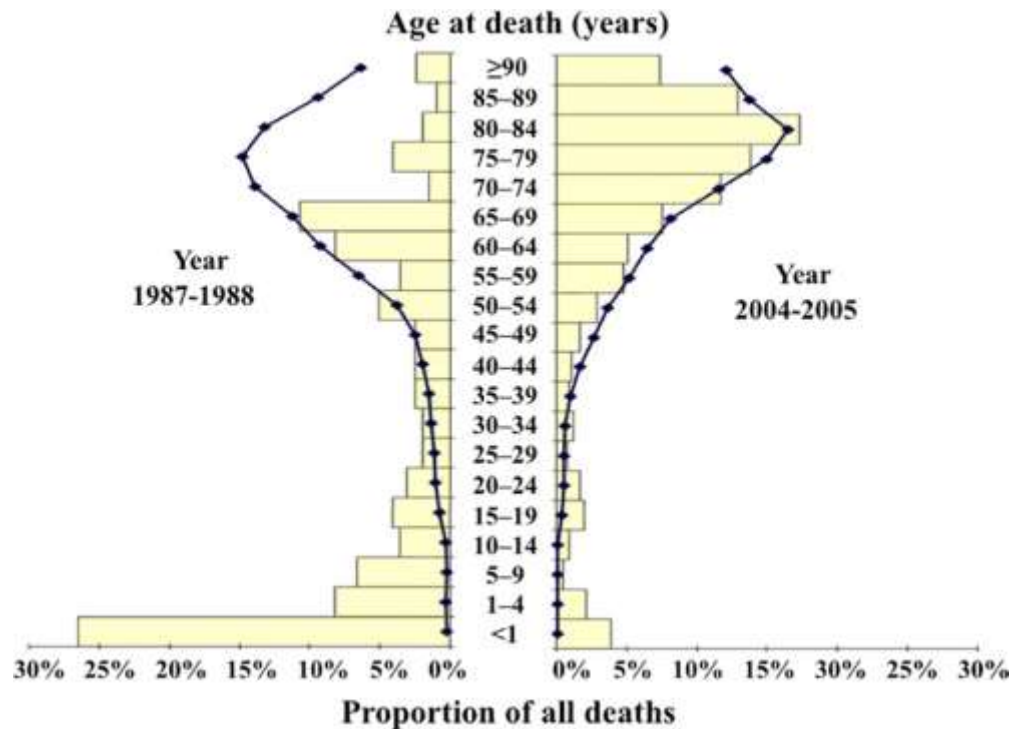
CHALLENGE is funded by Hellenic Society of Cardiology



# Changing demographics of CHD

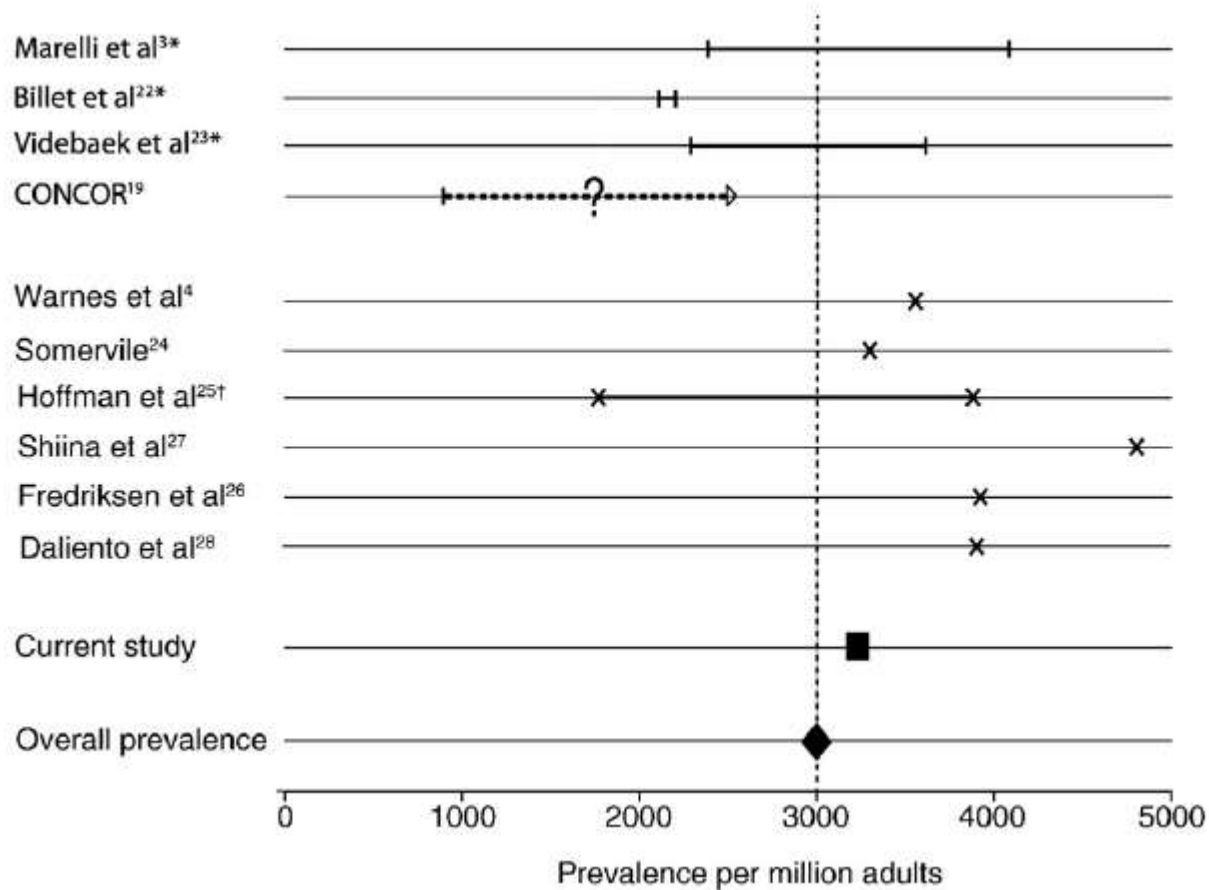


# Changing mortality in ACHD

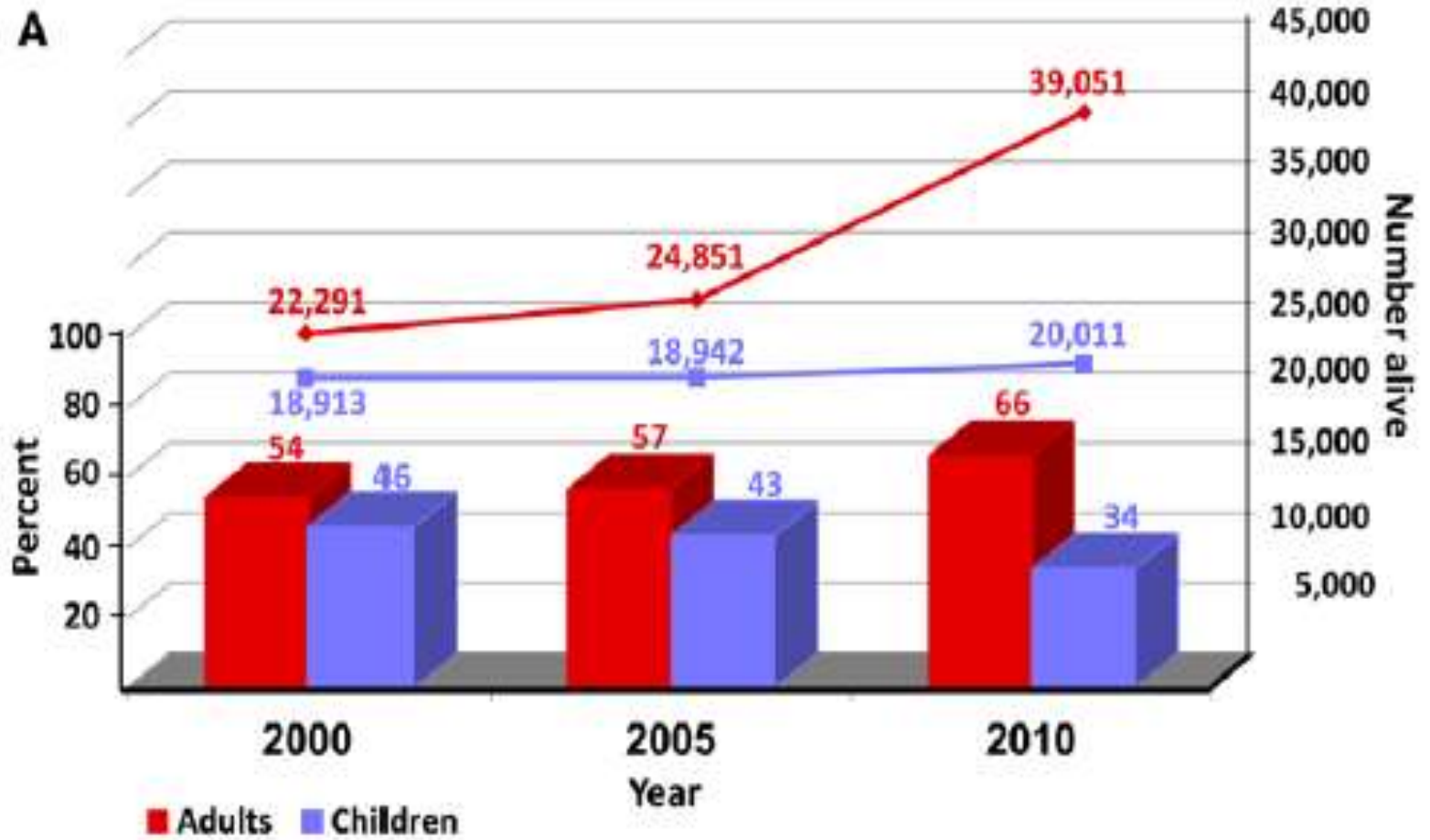


# The prevalence of adult congenital heart disease, results from a systematic review and evidence based calculation

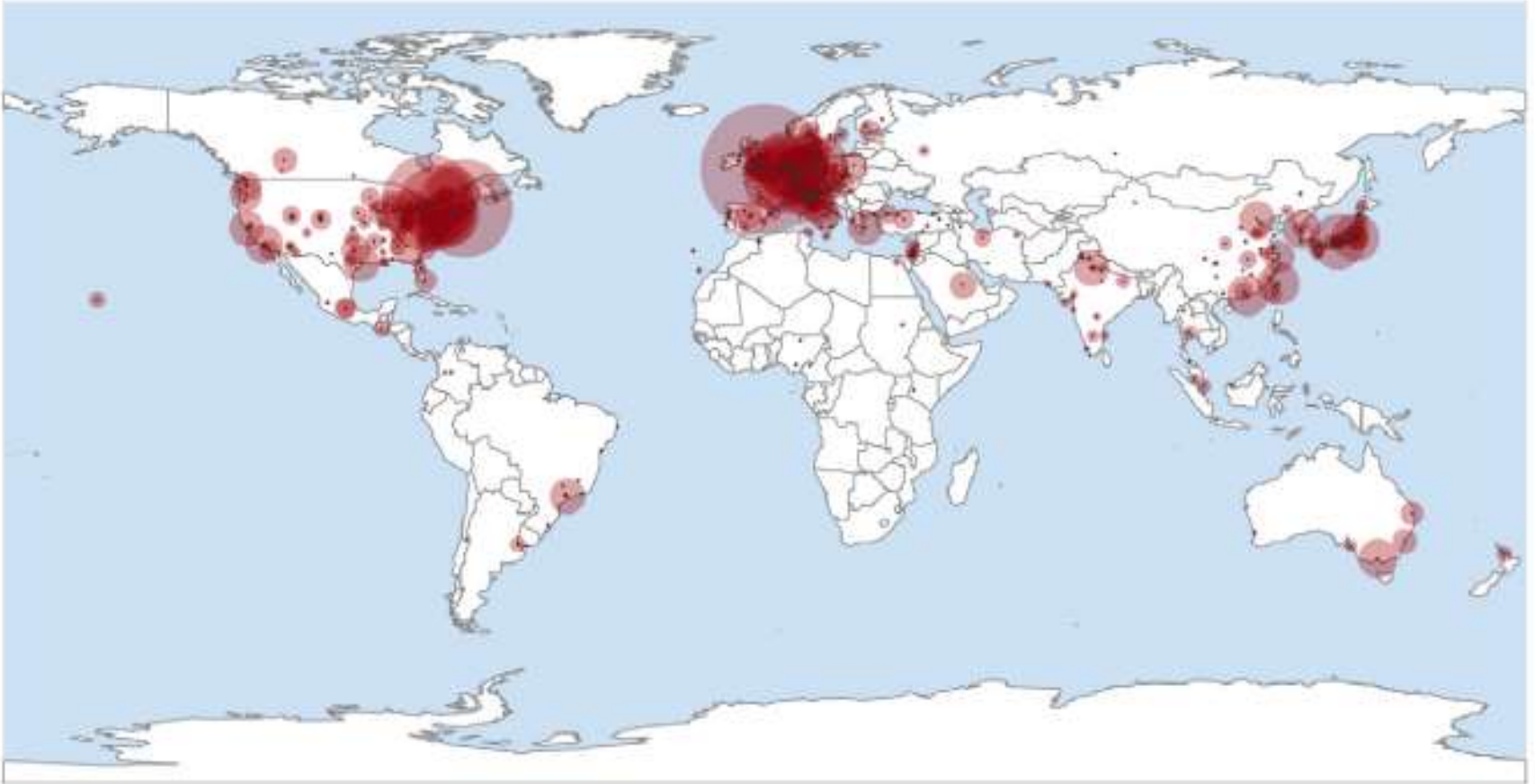
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The number of adults with CHD exceeds the number of children by **2:1**



# ACHD centers worldwide



# Background

Up to now there are no data on the epidemiology of CHD in Greece





# Purpose

- Estimation of the prevalence and long-term outcomes of congenital heart defects in Greece
- Development of an efficient organizational structure for the improvement of healthcare for patients with congenital heart disease

# Methods and Materials

- Initiation of the registry in January 2012
- Informed consent
- Patients >16 years old
- 18 expert ACHD centers

# Methods and Materials



Αρχική σελίδα
Εισαγωγή νέου ασθενούς
Επόμενη επίσκεψη ασθενούς
Αναζήτηση ασθενούς
Αλλαγή κωδικού
Βοήθεια
Αποσύνδεση

**Εισαγωγή**

Παρακαλώ εισάγετε τα στοιχεία για την αρχική επίσκεψη του νέου ασθενούς παρακάτω :

• Αριθμός ασθενούς :  • Ημερομηνία Επίσκεψης :  • Φύλο :  APPEN  ΘΗΛΥ

• Όνομα :  • Επώνυμο :  Πατρώνυμο :  • Ημερομηνία Γέννησης :

• Εισαγωγή Κύριας Διάγνωσης :  • Έτος Κύριας Διάγνωσης :

Αριθμός χειρουργικών επεμβάσεων :  Αριθμός λοιπών επεμβατικών πράξεων :

Λήψη φαρμάκων:  Αντιπηκτικά:  Καρδιακής ανεπάρκειας:   
 Ανταρρυθμικά:  Αντιυπερτασικά:  Ανταμοιοπεταλιακά:   
 Φάρμακα ΠΑΥ:

• Λειτουργικό στάδιο:  I  II  III  IV Ημερομηνία Επόμενης Επίσκεψης:

Επιπρόσθετες Διαγνώσεις :

Επιλέξτε από τις λίστες :  +

Παρεμβάσεις :

Επιλέξτε από την λίστα :  +

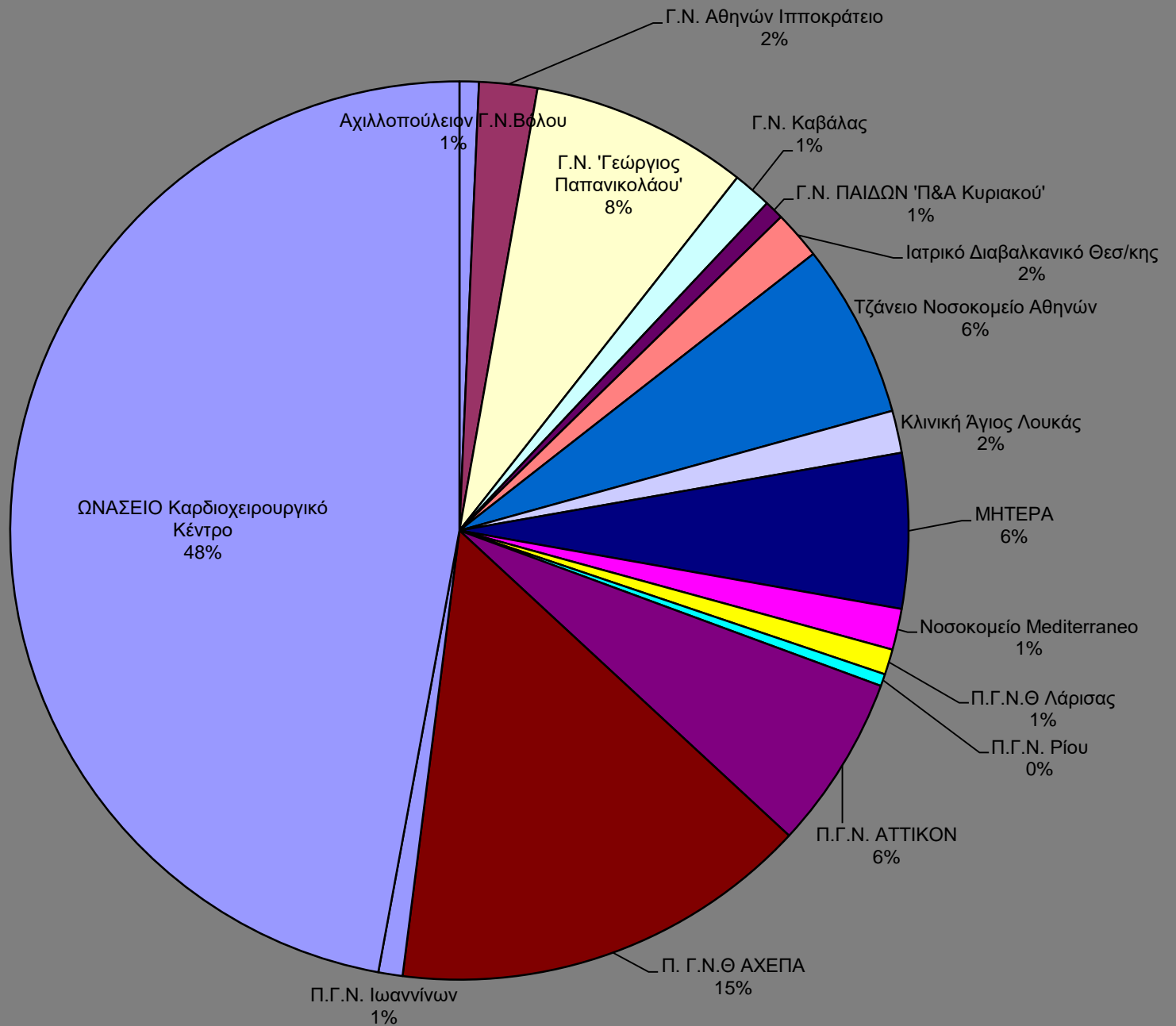
Σχόλια :

**Καθαρισμός** **Υποβολή**

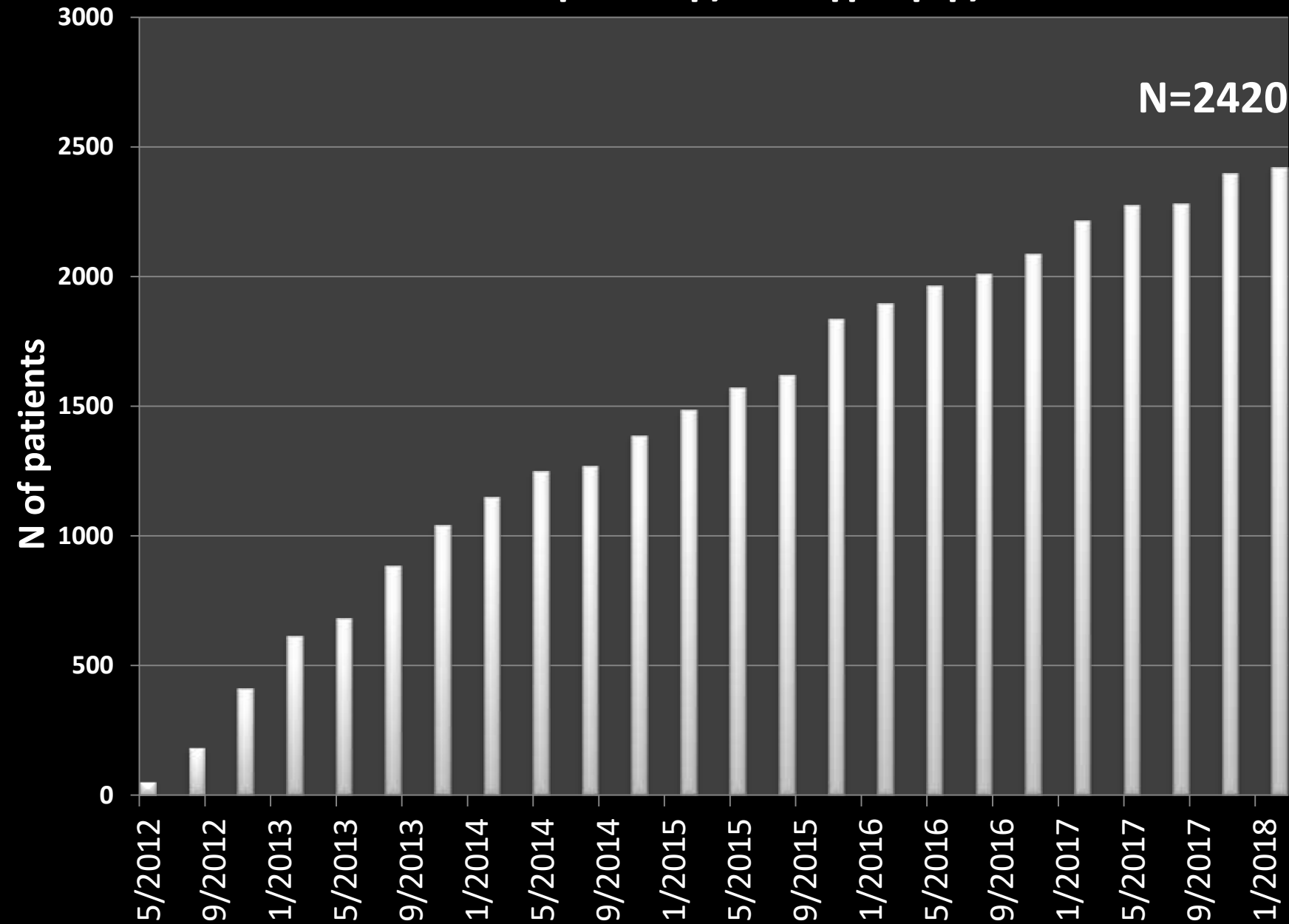
# Results

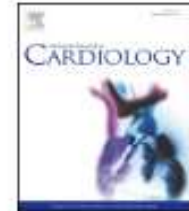


# ACHD centers



# Πορεία της καταγραφής





## Adult congenital heart disease in Greece: Preliminary data from the CHALLENGE registry



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### ABSTRACT

**Background:** The majority of patients with congenital heart disease (CHD), nowadays, survives into adulthood and is faced with long-term complications. We aimed to study the basic demographic and clinical characteristics of adult patients with congenital heart disease (ACHD) in Greece.

**Methods:** A registry named CHALLENGE (Adult Congenital Heart Disease Registry. A registry from Hellenic Cardiology Society) was initiated in January 2012. Patients with structural CHD older than 16 years old were enrolled by 16 specialized centers nationwide.

**Results:** Out of a population of 2115 patients with ACHD, who have been registered, (mean age 38 years (SD 16), 52% women), 47% were classified as suffering from mild, 37% from moderate and 15% from severe ACHD. Atrial septal defect (ASD) was the most prevalent diagnosis (33%). The vast majority of ACHD patients (92%) was asymptomatic or mildly symptomatic (NYHA class I/II). The most symptomatic patients were suffering from an ASD, most often the elderly or those under targeted therapy for pulmonary arterial hypertension. Elderly patients (>60 years old) accounted for 12% of the ACHD population. Half of patients had undergone at least one open-heart surgery, while 39% were under cardiac medications (15% under antiarrhythmic drugs, 16% under anticoagulants, 16% under medications for heart failure and 4% under targeted therapy for pulmonary arterial hypertension).

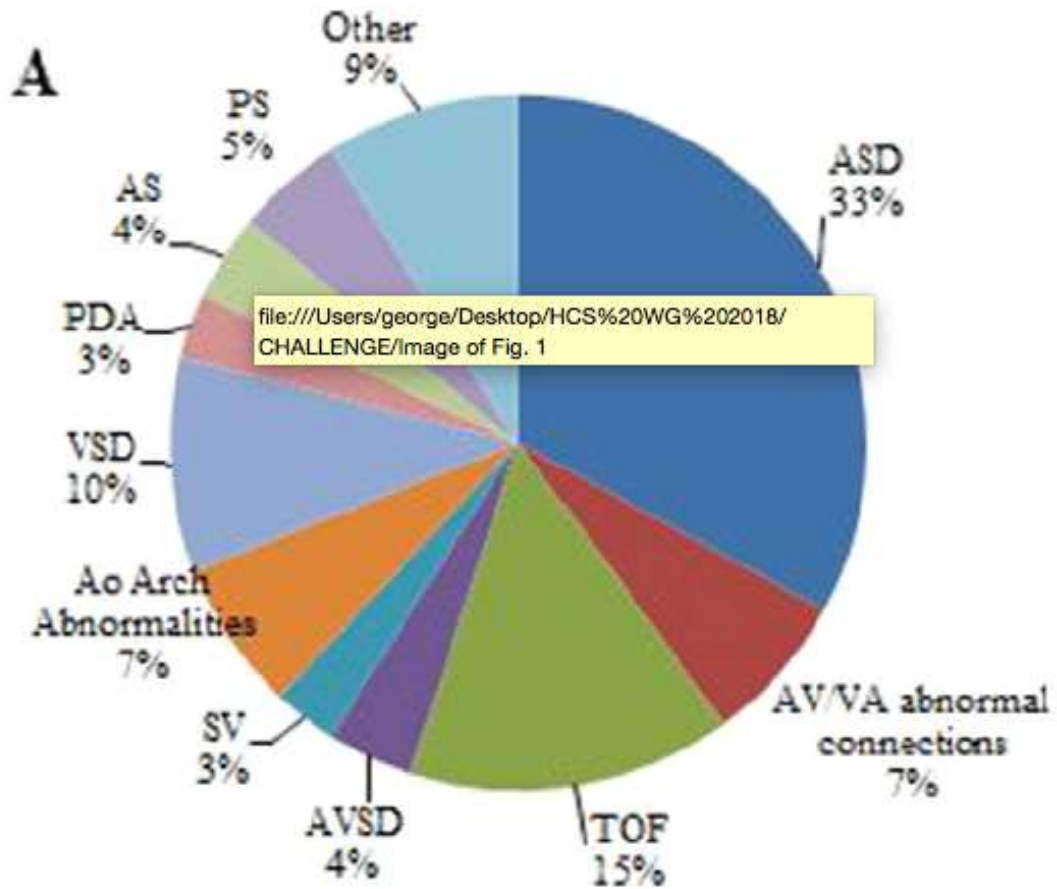
**Conclusions:** ACHD patients are an emerging patient population and national prospective registries such as CHALLENGE are of unique importance in order to identify the ongoing needs of these patients and match them with the appropriate resource allocation.

# Results until April 2017

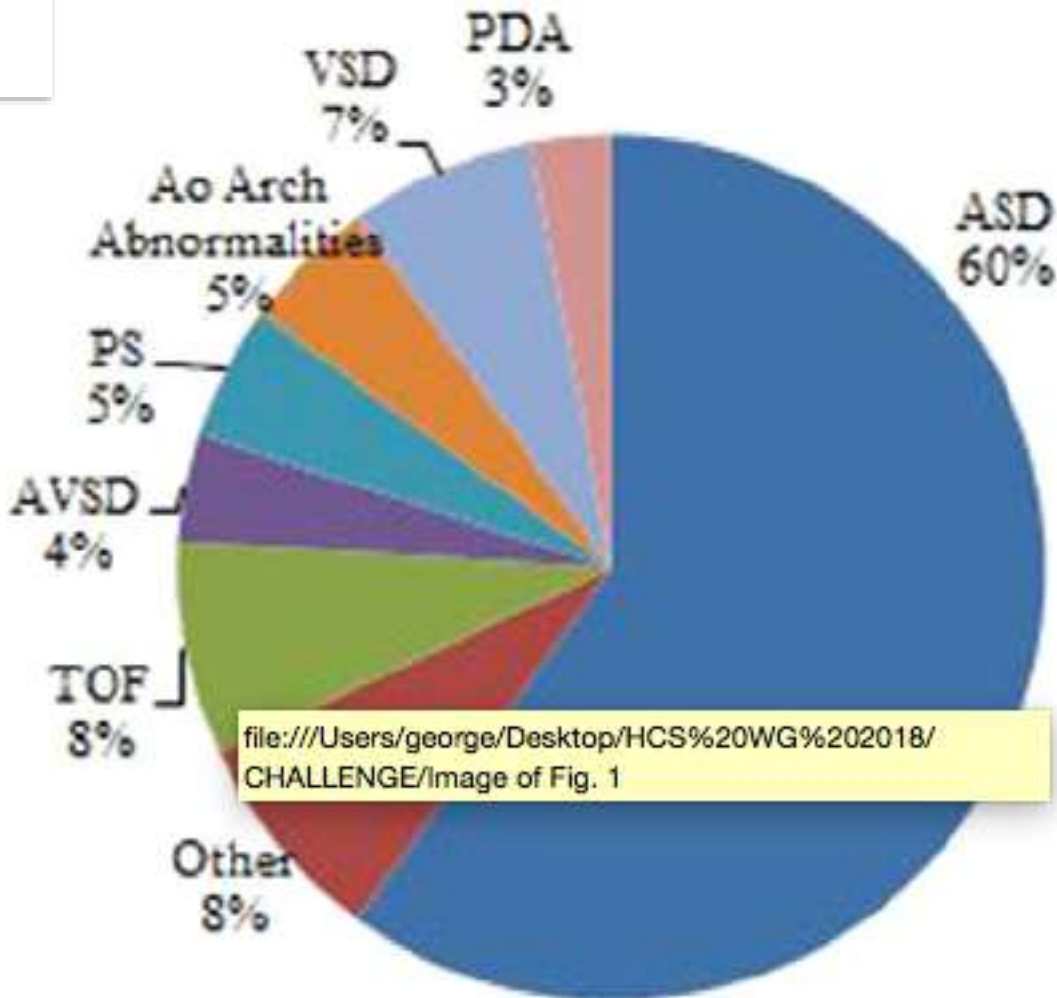
- N=2115
- mean age 38 years (SD 16)
- 52% women
- 47%-> mild, 37%-> moderate, 15%-> severe ACHD
- 12% elderly (>60 years)



# ACHD subtypes



# Elderly = 12% of the population



More often **mild CHD** and underwent **fewer surgeries**, while almost **two thirds** did not undergo any surgical procedure at all.

Only 38% of elderly patients were asymptomatic

# Results

**Table 1**  
Differences in epidemiological data according to the severity of CHD.

	Overall n = 2115	Mild n = 1008	Moderate n = 790	Severe n = 317	p-value
Age	38.0 ± 16.2	39.3 ± 17.3	37.7 ± 15.2	34.6 ± 13.9	<0.001
Males	1008 (47.7%)	438 (43.5%)	417 (52.8%)	153 (48.3%)	<0.001
NYHA I/II	1952 (92.3%)	965 (95.7%)	744 (94.2%)	243 (76.7%)	<0.001
NrSurgeries					
0	1061 (50.2%)	745 (73.9%)	232 (29.4%)	84 (26.5%)	<0.001
1	657 (31.1%)	212 (21.0%)	344 (43.5%)	101 (31.9%)	
≥2	397 (18.8%)	51 (5.1%)	214 (27.1%)	132 (41.6%)	
Cardiac Medication Use	821 (38.8%)	311 (30.9%)	315 (39.9%)	195 (61.5%)	<0.001
Antiarrhythmic drugs	314 (14.8%)	97 (9.6%)	137 (17.3%)	80 (25.2%)	<0.001
Medication for HF	342 (16.2%)	109 (10.8%)	132 (16.7%)	101 (31.9%)	<0.001
Anticoagulants	330 (15.6%)	138 (13.7%)	92 (11.6%)	100 (31.5%)	<0.001

Nr: Number; NYHA: New York Heart Association, HF: Heart Failure.

- 71% WHO I/II
- NYHA III/IV mainly ASD, TOF, AV/VA abnormal connection, single ventricle

# Results

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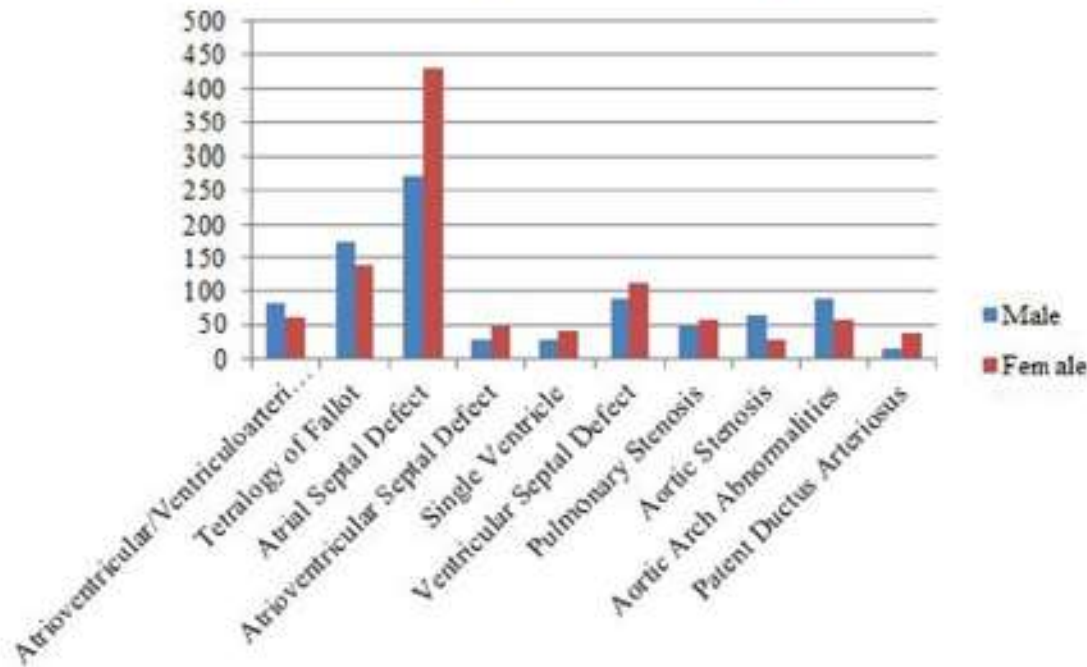
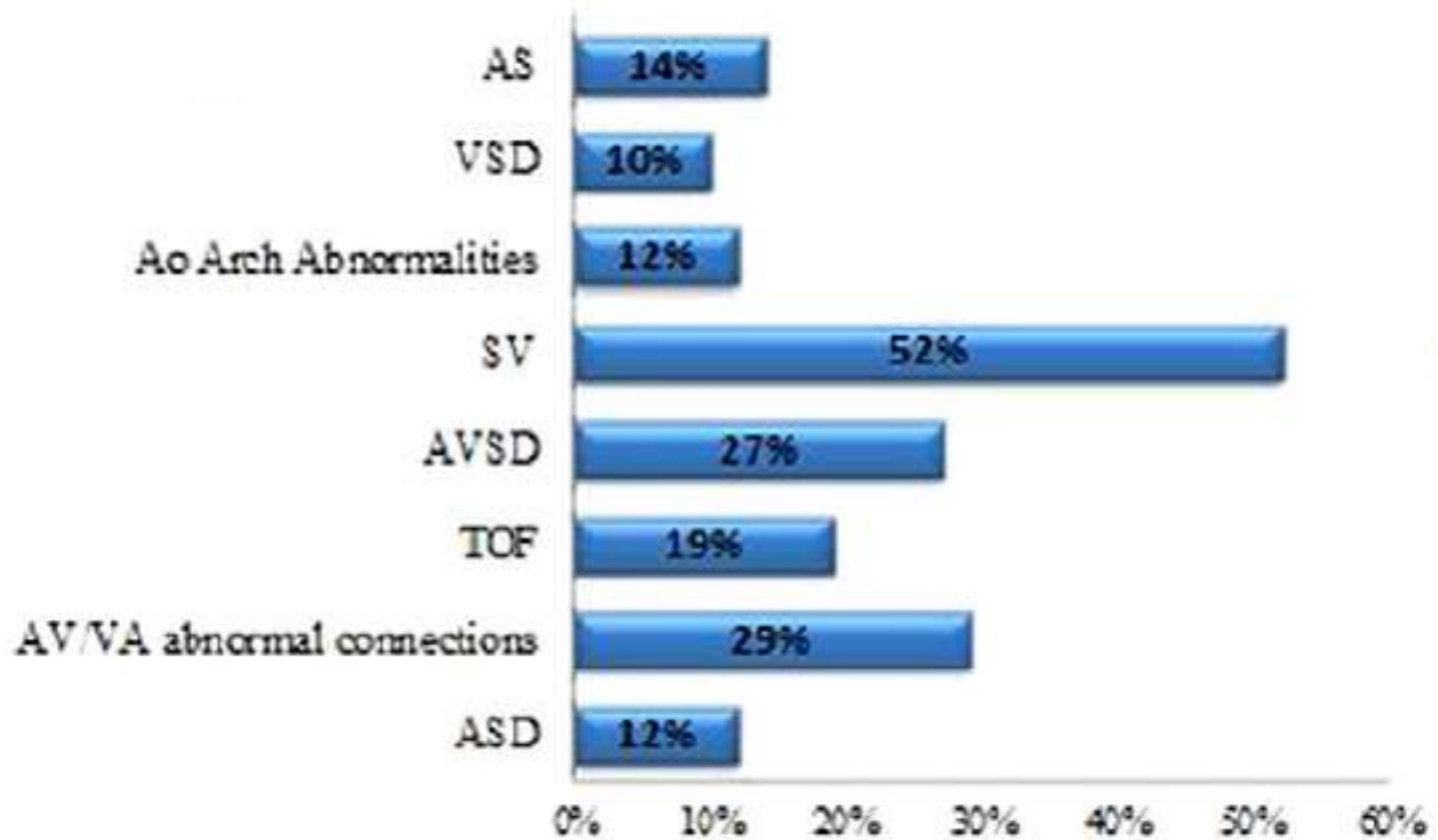


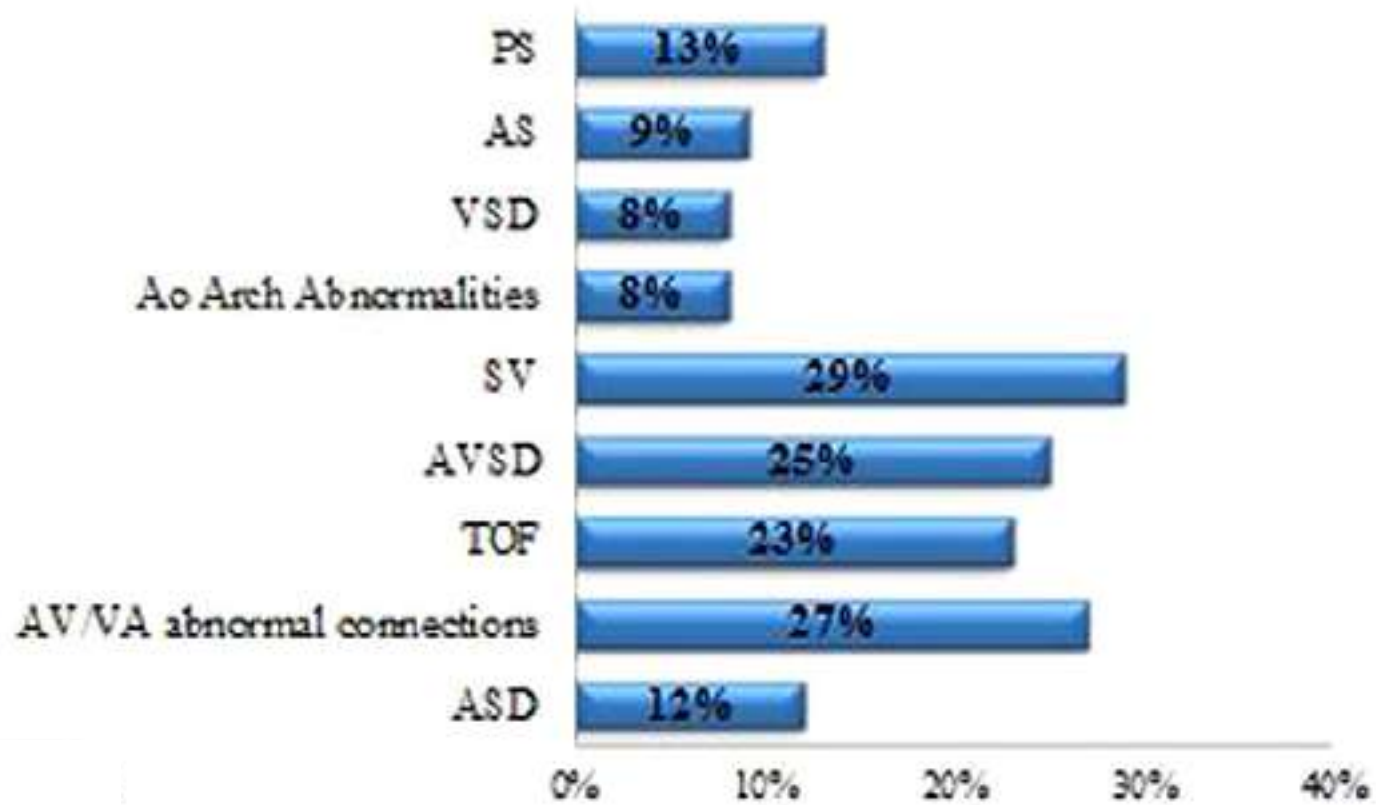
Fig. 2. The distribution of ACHD subtypes according to gender.

Females were **older** (mean age 40 years (SD 17) vs 36 years (SD 16),  $p < 0.001$ ), suffered more often from **mild** ACHD (52% vs 44%,  $p < 0.001$ ) and underwent **fewer surgeries** (no surgery was performed in 56% vs 44%,  $p < 0.001$ ).

# Heart failure in ACHD



# Antiarrhythmic medical treatment in ACHD





# Pulmonary hypertension in ACHD



Out of 2399, **N=68** are in PAH treatment ( $\approx 3\%$ )

**8 excluded:**  
3 Fontan  
5 Segmental PAH

Eisenmenger  
**42 pts (70%)**

L-> R  
**12 pts (20%)**

Small defects  
**1 pt (2%)**

After repair  
**6 pts (10%)**

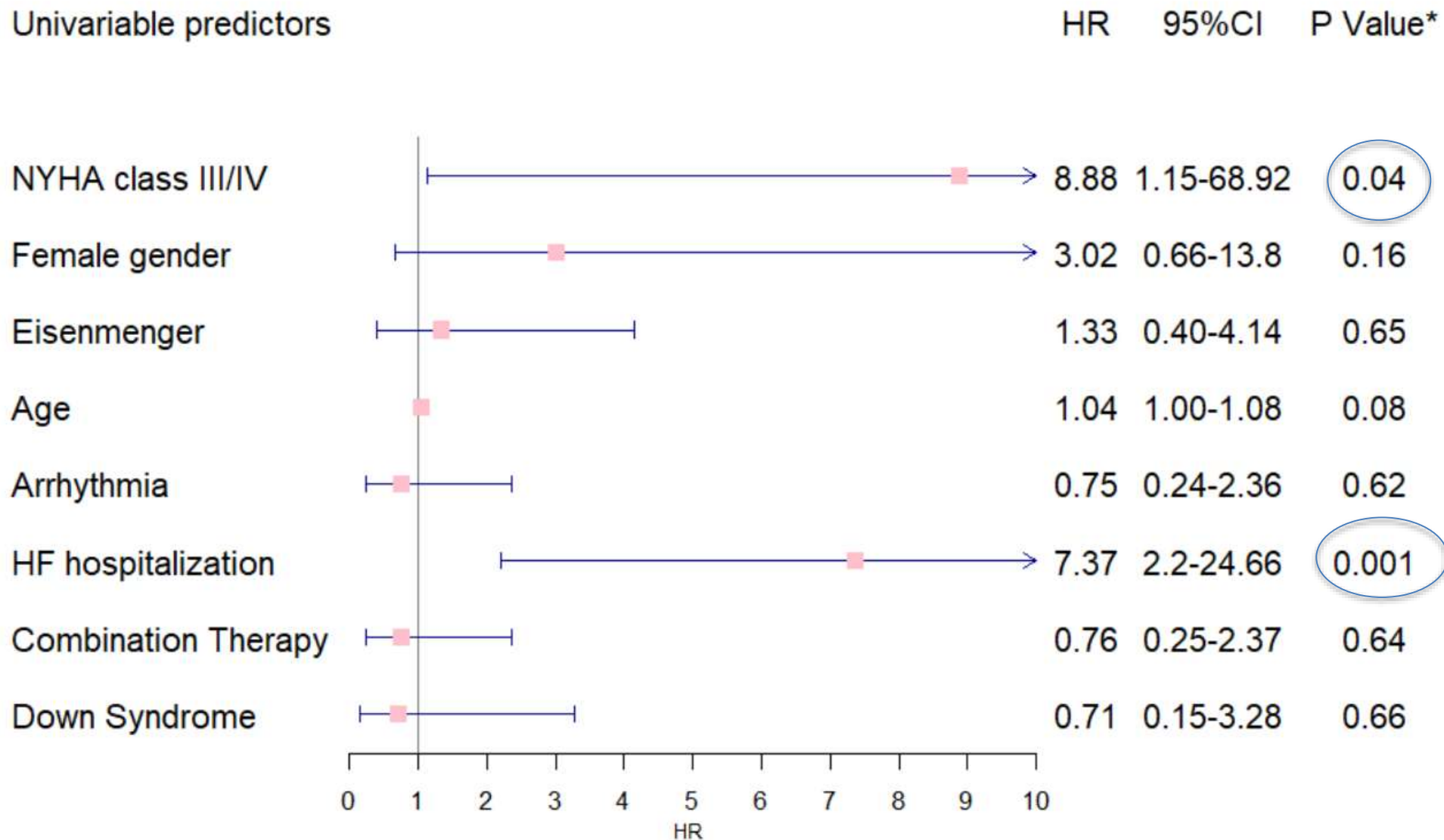
# Baseline Characteristics (n=60)

<b>Age</b>	46.5±14.8
<b>Males</b>	21 (35%)
<b>NYHA III/IV</b>	33 (55%)
<b>Down Syndrome</b>	9 (15%)
<b>PAH pharmacotherapy</b>	
ERA	54 (90%)
PDE5i	29 (48%)
Prostanoids	4 (7%)
<b>Combination therapy</b>	26 (43%)
<b>Oxygen therapy</b>	13 (22%)

# Complications

<b>Arrhythmias</b>	19 (32%)
<b>Endocarditis</b>	1 (2%)
<b>Stroke</b>	3 (5%)
<b>Pulmonary thrombosis</b>	4 (7%)
<b>HF hospitalization</b>	15 (25%)
<b>Hemoptysis</b>	2 (3%)
<b>Death</b>	12 (20%)
RHF	6 (10%)
Sepsis	2 (3%)
Arrhythmia	1 (2%)
Sudden death	1 (2%)
Massive pulmonary hemorrhage	1 (2%)
Unknown	1 (2%)

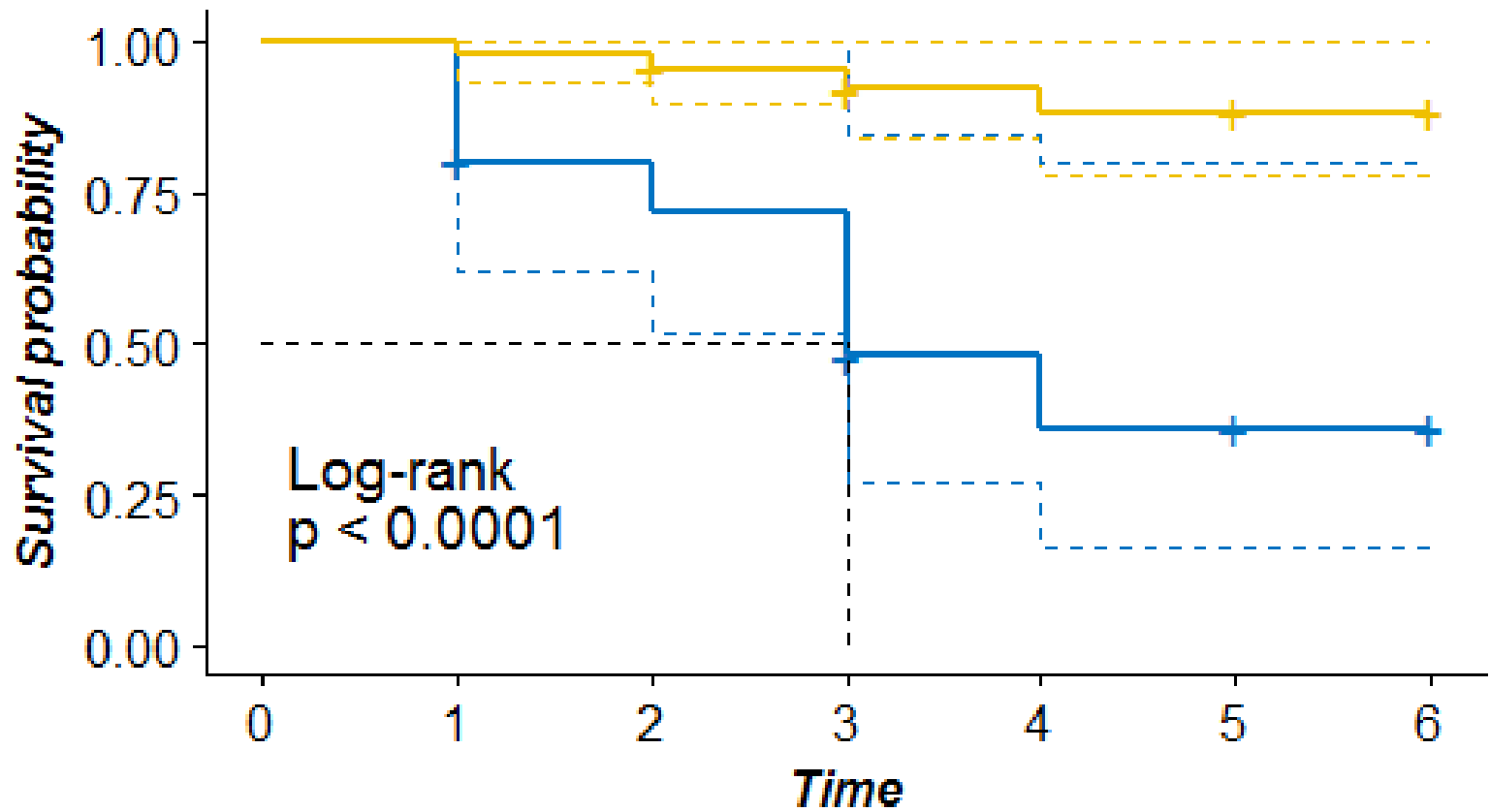
## Univariable predictors for cumulative mortality risk



\*Based on Univariate Cox model

# Survival Curves

Survival + HF Hospitalized + Not HF hospitalized



# Future perspectives->Follow up



Νέα επίσκεψη

Παρακαλώ εισάγετε τα στοιχεία για την επίσκεψη του ασθενούς παρακάτω :

• Λειτουργικό στάδιο:  I  II  III  IV

• Ημερομηνία Επίσκεψης:  Ημερομηνία Επόμενης Επίσκεψης:

Εγχείρηση:  Ημερομηνία:

Επεμβατική αντιμετώπιση:  Ημερομηνία:

Defibrillator:  Ημερομηνία:

Pace maker:  Ημερομηνία:

Αρρυθμίες / νοσηλεία:  Ημερομηνία:

Θάνατος:  Ημερομηνία:

Επιρόσθετες Διαγνώσεις :

Επιλέξτε από τις λίστες:



Παρεμβάσεις :

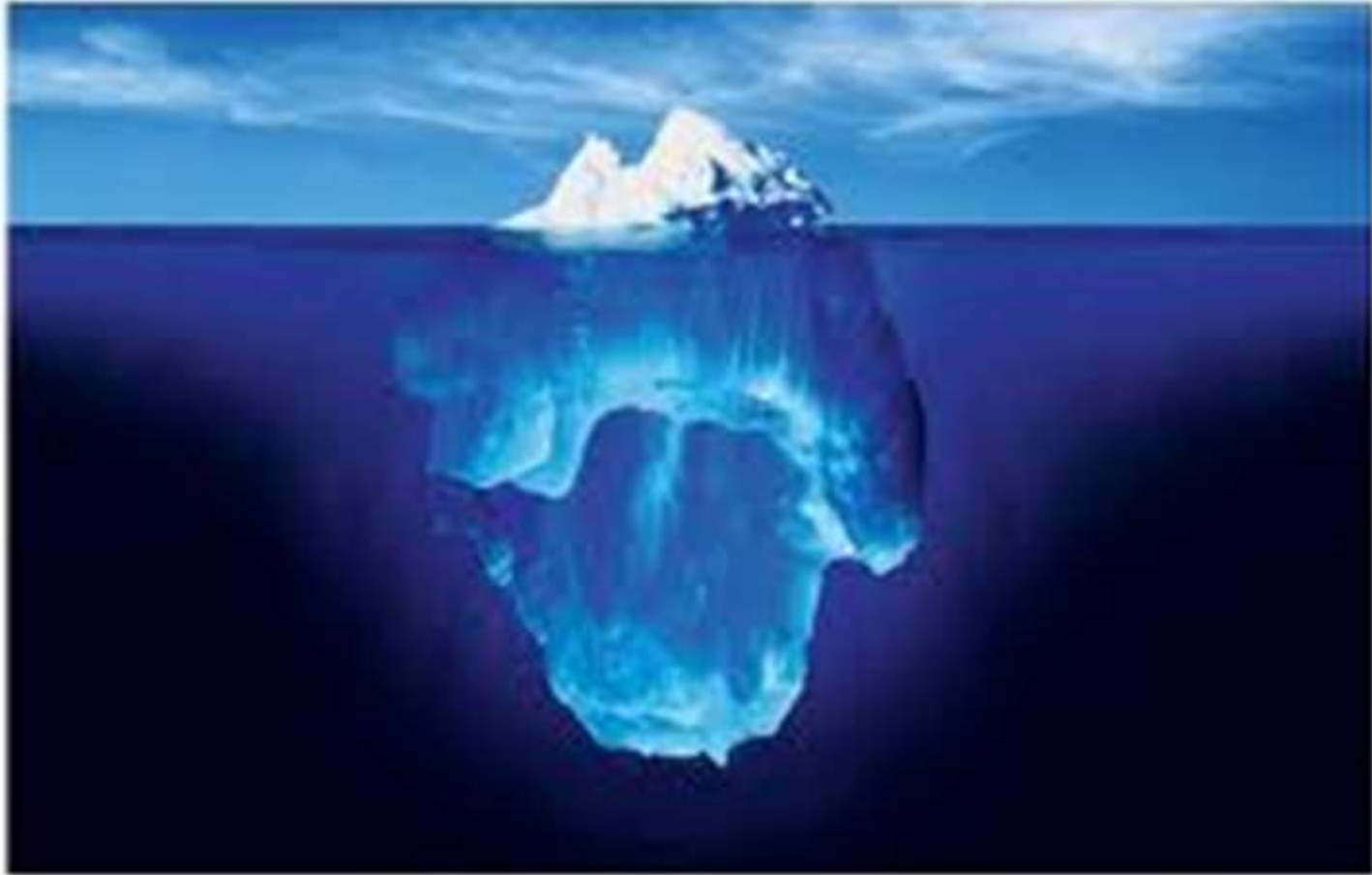
Επιλέξτε από την λίστα:



Καθαρισμός

Υποβολή

27600 ACHD patients wanted!!



# THE CHALLENGE TEAM

## Κέντρα ΣΚ:

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Α. Φρογουδάκη  
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Δ. Ντέλλος  
Σ. Μπρίλη  
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Α. Μακγίνας  
Μ. Παπαφυλακτού  
Ν. Καμπουρίδης  
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Π. Λαθρίδου  
Α. Πίτσης  
Δ. Ντιλούδη  
Ι. Κανακάκης  
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## Συμβάλλοντες στην καταγραφή

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Γ. Παρχαρίδης  
Κ. Βανταλής  
Ι. Βλασερός

## Νέα κέντρα

Α. Πατριανάκος  
Μ. Αργυρίου





Back Up

# Conclusions (I)

Severe ACHD patients:

- were younger
- more symptomatic
- underwent multiple surgeries
- were more frequently under medical therapy for heart failure and arrhythmias.

.

## Conclusions (II)

The most common form of ACHD that required HF treatment was SV.

Patients with SV, AV/ VA abnormal connections, TOF and AVSD were more frequently under treatment for arrhythmias.

# More to come....

- More registered patients
- More participating centers
- More investigators
- More results to publish



# Conclusions (III)

# Limitations

- Misclassification bias
- The entire data of the patients were not accessible
- The high proportion of expert centers might induce bias

