

**BAV: insights from a large,  
worldwide registry**

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**EUROVALVE 2021**

# CONFLICTS:

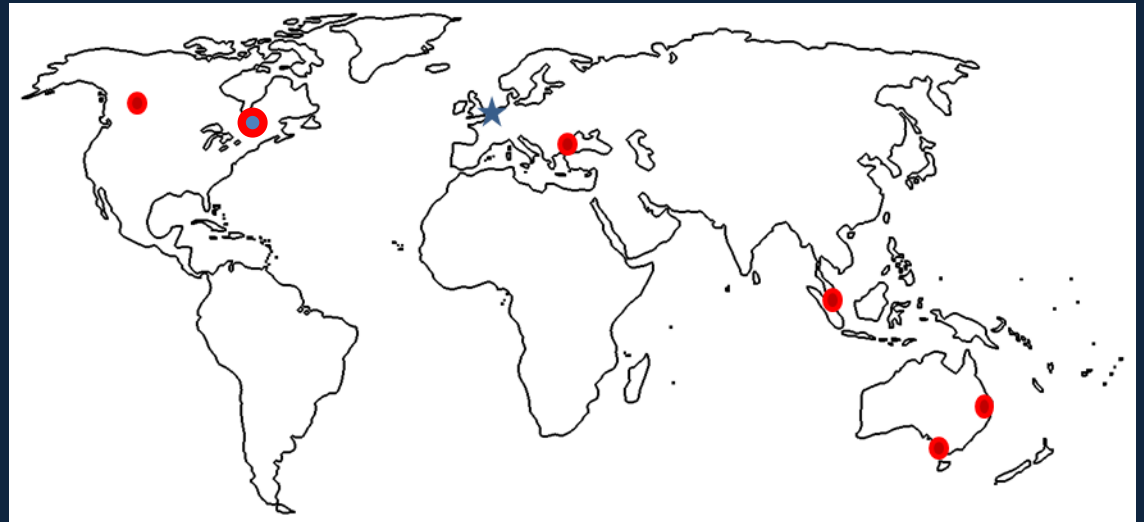
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Medical Center, The Netherlands

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Edwards Lifesciences, Abbott, GE Healthcare,  
Bayer, Novartis

# BAV International Registry

- N = 3075
- Origin:
  - The Netherlands
  - Singapore
  - Australia
  - Canada
  - Romania



- Systematic analysis of echocardiograms (not simple tabulation of clinical reports)

# BAV International Registry

- Characterization of the phenotype of BAV and prognostic implications
- Inter-ethnic differences in aortic root dimensions and aortic valve dysfunction
- Gender differences in BAV

# Prognostic Implications of Raphe in Bicuspid Aortic Valve Anatomy

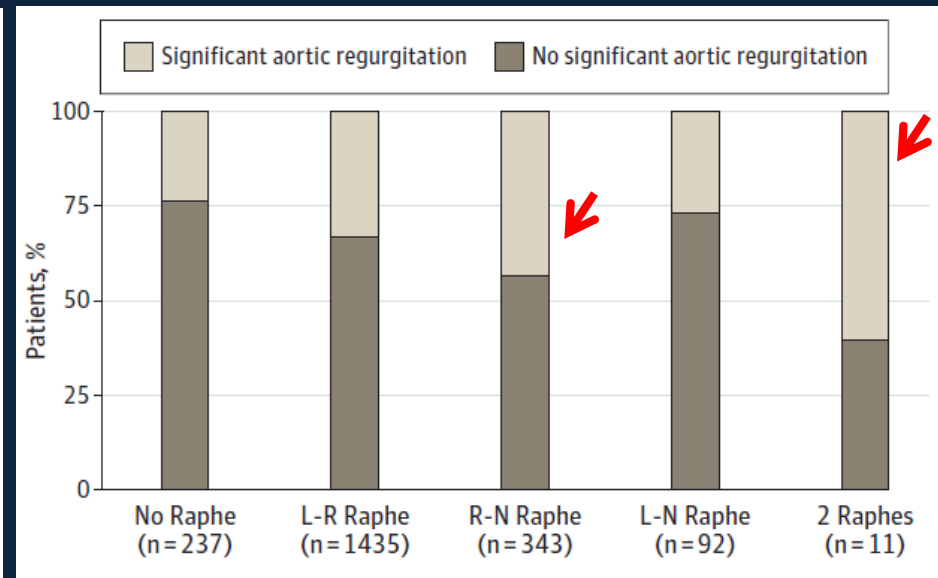
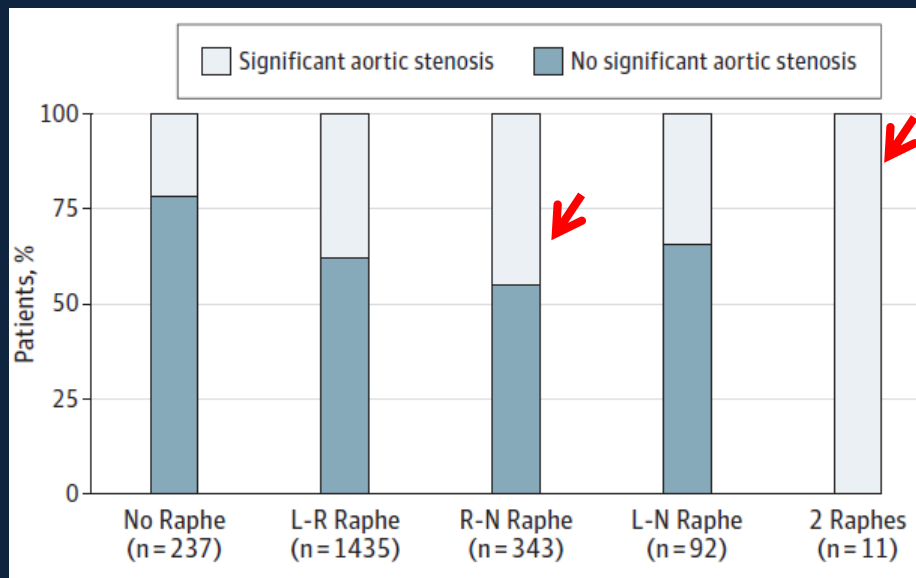
William K. F. Kong, MD; Victoria Delgado, MD, PhD; Kian Keong Poh, MD; Madelien V. Regeer, MD; Arnold C. T. Ng, MBBS, PhD; Louise McCormack, MBBS; Tiong Cheng Yeo, MD; Miriam Shanks, MD, PhD; Sarah Parent, MD; Roxana Enache, MD, PhD; Bogdan A. Popescu, MD, PhD; Michael Liang, MD; James W. Yip, MD; Lawrence C. W. Ma, MBBS; Vasileios Kamperidis, MD, PhD; Philippe J. van Rosendael, MD; Enno T. van der Velde, PhD; Nina Ajmone Marsan, MD, PhD; Jeroen J. Bax, MD, PhD

- N = 2118
- Mean age 47 years
- 72% male
  
- 89% BAV-raphe
- 11% BAV-no raphe

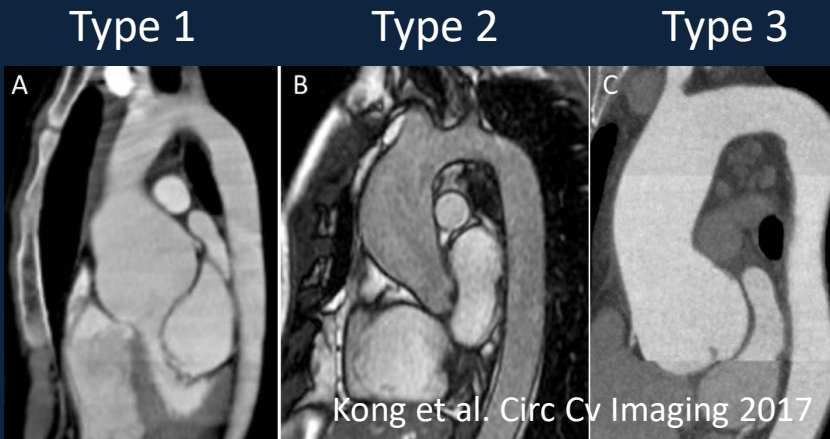
# Phenotype of BAV vs. valve function

**Moderate and severe  
aortic stenosis  
N = 779 (37%)**

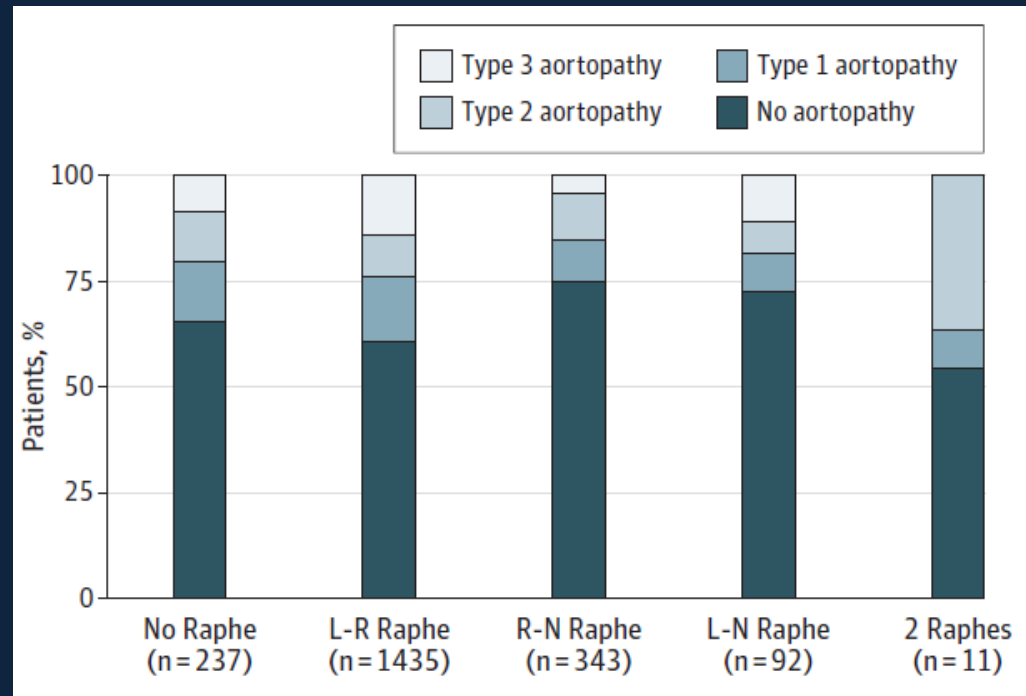
**Moderate and severe  
aortic regurgitation  
N = 679 (32%)**



# Phenotype of BAV vs. aortopathy



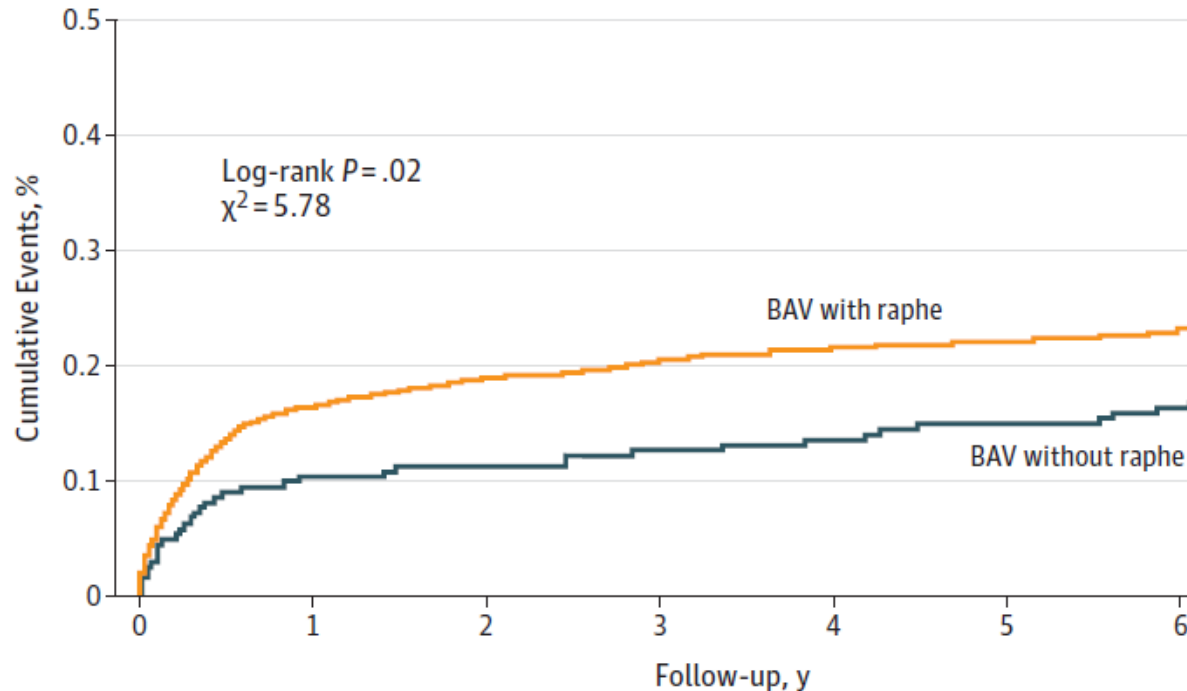
Type 1 = ao root  
 Type 2 = asc aorta  
 Type 3 = both



Types more/less equally distributed  
 Dissections not frequent but ..  
 patients with **aortic dissection** most often fusion **raphe between the left and right coronary cusps** (n=5, 63%), followed by fusion raphe between the right and the non-coronary cusps (n=2, 25%) and no raphe at all (n=1, 12%).

# Phenotype of BAV vs. AVR

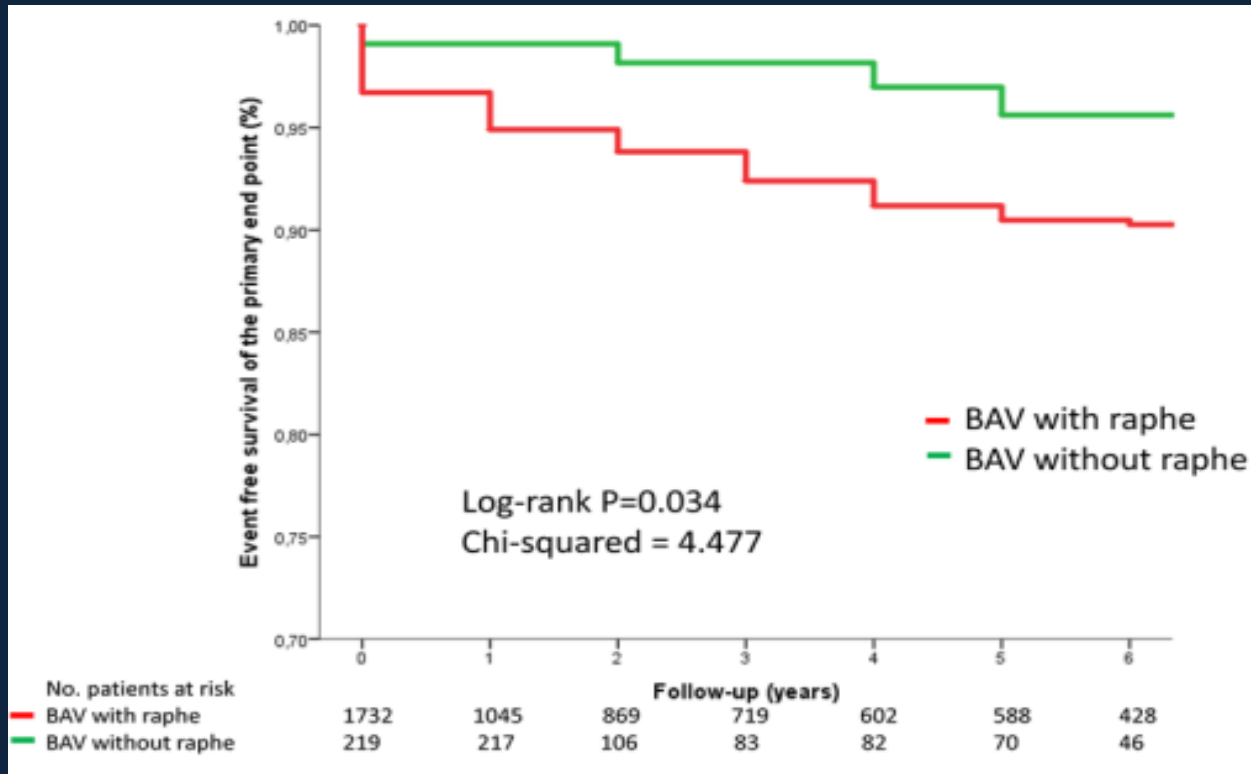
During a median follow-up of 2 years, 591 patients underwent AVR and 307 received aortic surgery



No. patients at risk	0	1	2	3	4	5	6
BAV with raphe	1782	1482	1437	1409	1390	1380	1362
BAV without raphe	221	198	196	192	190	187	184



# Phenotype of BAV vs. all-cause death



On multivariable Cox regression analysis, BAV with raphe was not independently associated with all-cause mortality



European Society  
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**CLINICAL RESEARCH**

*Valvular heart disease*

## **Inter-ethnic differences in valve morphology, valvular dysfunction, and aortopathy between Asian and European patients with bicuspid aortic valve**

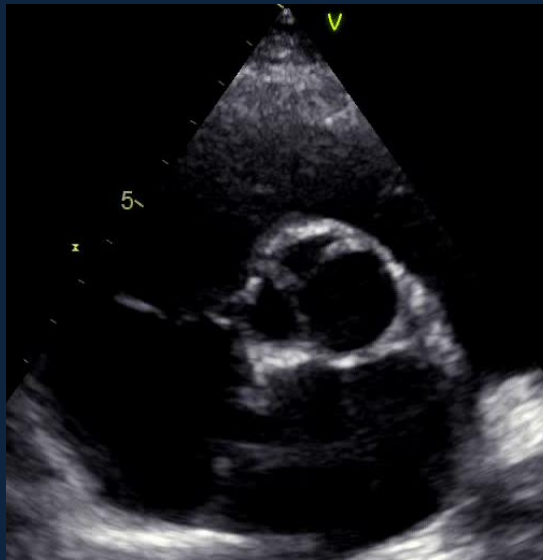
**William K. F. Kong<sup>1,2</sup>, Madelien V. Regeer<sup>1</sup>, Kian K. Poh<sup>2</sup>, James W. Yip<sup>2</sup>, Philippe J. van Rosendael<sup>1</sup>, Tiong C. Yeo<sup>2</sup>, Edgar Tay<sup>2</sup>, Vasileios Kamperidis<sup>1,3</sup>, Enno T. van der Velde<sup>1</sup>, Bart Mertens<sup>4</sup>, Nina Ajmone Marsan<sup>1</sup>, Victoria Delgado<sup>1</sup>, and Jeroen J. Bax<sup>1\*</sup>**

- **N = 1427**
  - 794 Europeans (mean age 44 years, 71% male)
  - 633 Asians (mean age 46 years, 72% male)

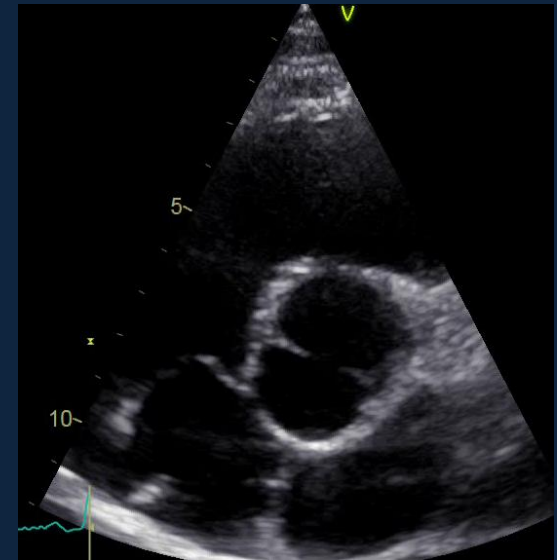
# Ethnicity vs. BAV type



**BAV**  
**left-right fusion**  
**68% Europeans**  
**67% Asians**



**BAV**  
**Non-right fusion**  
**More frequent**  
**in Asians**  
**19.7% vs. 13.6%**

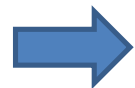


**BAV**  
**No raphe**  
**More frequent**  
**in Europeans**  
**14.5% vs. 6.8%**

# Ethnicity vs. Aortic dimensions

ALL PATIENTS:  
 70% normal  
 10% root only  
 10% ascending aorta only  
 12% diffuse dilated aorta

	All Patients (n = 1427)	European (n = 794)	Asian (n = 633)	P-value
Aortic dimensions				
Aortic annulus indexed diameter (mm/m <sup>2</sup> )	12.6 ± 1.9	12.2 ± 1.8	13.2 ± 1.9	<0.001
Sinus of Valsalva indexed diameter (mm/m <sup>2</sup> )	18.7 ± 3.6	18.1 ± 3.3	19.6 ± 3.7	<0.001
Sinotubular junction indexed diameter (mm/m <sup>2</sup> )	15.7 ± 3.4	15.6 ± 3.5	15.9 ± 3.2	<0.001
Ascending aorta indexed diameter (mm/m <sup>2</sup> )	20.1 ± 4.2	19.7 ± 4.0	20.7 ± 4.5	<0.001
Pattern of aortic dilation				0.008
Normal	987 (69.2)	538 (67.8)	449 (70.9)	
Dilated aortic root only	133 (9.3)	73 (9.2)	60 (9.5)	
Dilated ascending aorta only	138 (9.7)	69 (8.7)	69 (10.9)	
Diffuse dilation of aorta	169 (11.8)	114 (14.4)	55 (8.7)	



# Ethnicity vs. Aortic dimensions

Indexed dimensions of the aortic annulus, sinus of Valsalva, sinotubular junction and ascending aorta were significantly larger in Asians as compared to Europeans....  
Although Asians smaller than Europeans....

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ORIGINAL ARTICLE - VALVULAR HEART DISEASE VALVULAR HEART DISEASE

## **Sex Differences in Phenotypes of Bicuspid Aortic Valve and Aortopathy**

**Insights From a Large Multicenter, International Registry**

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**See Editorial by Michelena and Mankad**

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William K.F. Kong, MD, Madelien V. Regeer, MD, Arnold C.T. Ng, MBBS, PhD, Louise McCormack, MBBS, Kian Keong Poh, MD, Tiong Cheng Yeo, MD, Miriam Shanks, MD, PhD, Sarah Parent, MD, Roxana Enache, MD, PhD, Bogdan A. Popescu, MD, PhD, James W. Yip, MD, Lawrence Ma, MBBS, Vasileios Kamperidis, MD, PhD, Enno T. van der Velde, PhD, Bart Mertens, PhD, Nina Ajmone Marsan, MD, PhD, Victoria Delgado, MD, PhD, and Jeroen J. Bax, MD, PhD

- **N = 1992**
- **Men 1424, 71.5%**
- **Women 568, 28.5%**

# Analysis of the BAV morphology

## Sievert Classification

- Overall population:
- most common BAV morphology: type 1 with raphe between left and right cusps (68.7%)
- Least frequent BAV morphology: type 2 with 2 raphe (0.4%)
- No differences in BAV morphology between men and women

# Valve dysfunction

- 84% of patients have some degree of valve dysfunction
- Men more AR: 34.8% vs 22.2% ( $P < 0.001$ )  
related to more often dilated aorta/root
- Women more AS: 44.1% vs 34.5% ( $P < 0.001$ )

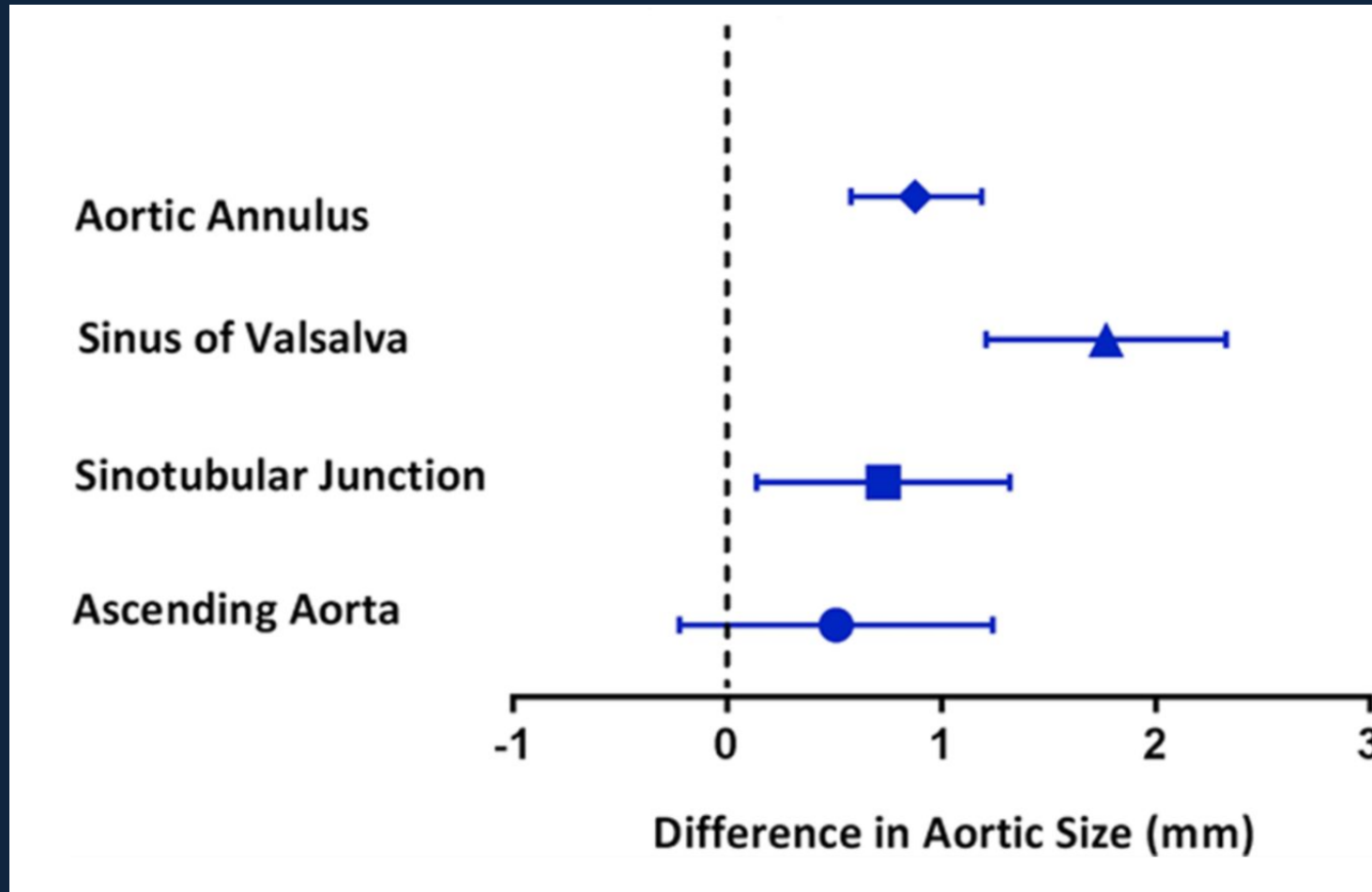


# Aortopathy in 697 patients, 35%

Men:

- more often isolated aortic root dilation (14.2% vs 6.7%,  $P < 0.001$ ) ( $\rightarrow$  AR)
- Diffuse aorta dilation (16.2% vs 7.3%,  $P < 0.001$ )

# Women: smaller aortic annulus, sinus of Valsalva and STJ versus men



# LARGE international registry

Provides information on:

Prevalence of BAV phenotypes and outcomes

Differences in BAV phenotypes and ethnicities

Differences in BAV and gender

# Presence of raphe in BAV

- Raphe in general: More frequently associated with significant valve dysfunction and aortic dilation
- The location of raphe: was associated with the type of valve dysfunction but not with aortic dilation
- Raphe in general: associated with increased risk of valve surgery and all-cause mortality

# Interethnic differences in BAV

- Asian patients have larger aortic annulus and root; this may have important implications for:
  - Follow-up of valve function and aortic dimensions
  - evolving technologies in aortic valve replacement (TAVR – larger size in Asian patients)
  - Indications for aortic surgery

# Gender differences in BAV

- Men more often AR and dilated root/asc
- Women more often AS
- Women: smaller aortic annulus, sinus of Valsalva and STJ versus men
- Implications for therapies