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以下文章来源于MiHeart，作者肥肥黄馒头



MiHeart

心脑血管领域的最新文献和视频，仅是个人业余时间的整理和分享，希望一起探...



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昨晚和一位老师聊了下二叶主动脉瓣，深觉自己之浅薄，所以今天斗胆再来写一写。

人类对心脏瓣膜的研究可追溯到文艺复兴-达芬奇的时代，其中，二叶主动脉瓣（Bicuspid aortic valve, BAV）很早即被观察到。在1961年的Circulation杂志中，病理学家 Jesse E. Edwards对其进行了第一次总结，可以仔细阅读下文章，发现哪些知识其实早在60年前即已获得。

Editorial

The Congenital Bicuspid Aortic Valve

IT IS commonly taught that the congenital bicuspid aortic valve may function as perfectly as does the normal tricuspid aortic valve. One may wonder, however, about the wisdom of this teaching when certain facts concerning the congenital bicuspid aortic valve are reviewed.

The congenital bicuspid aortic valve in its pure state, without the addition of acquired disease, may be grossly incompetent as proved by clinical and necropsy observations. For example, a congenital bicuspid valve occurs in about 85 per cent of patients who have coarctation of the aorta; Christensen and Hines¹ observed basal diastolic murmurs in 20 per cent of a clinical series of 96 patients with coarctation of the aorta.

Bacterial endocarditis, which tends to involve tissue subjected to trauma, has a distinct predilection for the congenital bicuspid aortic valve. In almost half of the cases of aortic valvular bacterial endocarditis, the underlying disease is a congenital bicuspid aortic valve.

Smith and Matthews² re-emphasized the views of Peacock, expressed in the last century, that the congenital bicuspid valve is particularly prone to become calcified and therefore stenotic. It is recognized that a form of congenital aortic stenosis exists but, in this condition, the valve is essentially dome-shaped and not bicuspid. This anomaly

is not the subject of this editorial, although the valve may acquire calcific change and in this way, perhaps, the degree of stenosis may become accentuated.

One must assume that the occurrence in a congenital bicuspid aortic valve of intrinsic insufficiency and the traumatic background leading to bacterial endocarditis results from improper closure of the valve. Furthermore, the tendency for congenital bicuspid aortic valves to calcify is perhaps also a manifestation of trauma related to improper closure. With this background suggesting that the congenital bicuspid aortic valve may close improperly, one may present some theoretic considerations on the relationship of function to the structure of the bicuspid aortic valve.

Consideration of the normal tricuspid aortic valve emphasizes how nearly perfect its structure is for its function. The three cusps of the normal aortic valve may be looked on as three independent units, each being connected to its respective segment of the aortic wall. Each cusp has two lateral attachments to the aortic wall and an inferior attachment to the root of the aorta. Between the two lateral attachments of a cusp the tissue is greater than the straight-line distance between the points of attachments. This extra length of tissue of a normal tricuspid aortic valve allows the central part of each cusp to extend to the center of the aortic orifice where it touches the other cusps during closure of the valve (fig. 1a). The excess

From the Section of Pathologic Anatomy, Mayo Clinic, Rochester, Minnesota.



图2 1985年，病理学家Jesse E. Edwards博士与心脏专家和医学生一起检查心脏标本（这位2008年96岁高龄的时候已经去世了，表示缅怀）

在1980年代的尸检报道中，研究者们即发现其最常累及右和左瓣叶，在先天性二叶瓣畸形中，左冠状动脉开口出现在窦管交界处或上方的比例为44%，而后天者中则不到20%。1989年Angelini教授的形态报道，进一步提示主动脉瓣作为瓣膜复合体，发育过程中大多数起源于三叶，部分在宫内发育过程或出生后变为二叶；且BAV的融合模式具有明显变异性。

Br Heart J 1981; **45**: 142-7

Coronary arterial anatomy in bicuspid aortic valve *Necropsy study of 100 hearts*

PAUL K LERER,* WILLIAM D EDWARDS

*From the Department of Pathology and Anatomy and from the Mayo Medical School,
Mayo Clinic and Foundation, Rochester, Minnesota, USA*

图3 1981年，对100例BAV患者冠脉解剖的尸检报道



图4 Gianni D. Angelini教授（以防你认不出来，这位还是不停跳心脏搭桥术的创始人，英国医学科学院院士）

基本上提到BAV在一般人群中的患病率，综述都会引用2000年前后的三篇文献写为：0.5-2%，此处笔者也不能免俗。然而，大家都有个既往印象，流行病学调查显示亚洲人群BAV的发病率是高于西方人群的，不过这个既往证据相对薄弱，也未分型。具体人群间的区别，则一直等到了2018年发表在EHJ上的一篇文章，欧洲研究者 William Kf Kong教授（这是位亚裔学者），针对西方和亚洲BAV人群的超声检查比较，欧洲人群794名：亚洲人群633名（主要是新加坡），其中亚洲人群高血压、血脂异常和糖尿病患病率高于欧洲组，但吸烟率较低。发现两组人群正常功能的BAV患病率相似（12.6% vs 12.3%， $p=0.936$ ），狭窄多于关闭不全。但BAV形态存在显著差异，均主要为1型（欧洲68.5% vs 亚洲 67.0%），然而欧洲人群更常观察到0型（14.5% vs 6.8%， $p<0.001$ ），而在亚洲人群1型中R-N则更多见（19.7% vs 13.6%， $p<0.001$ ）。这与2013年韩国研究的结果相似。（有意思的是，欧洲BAV人群的严重返流发生率高于亚洲人群，44.2% vs 26.8%， $p<0.001$ ）

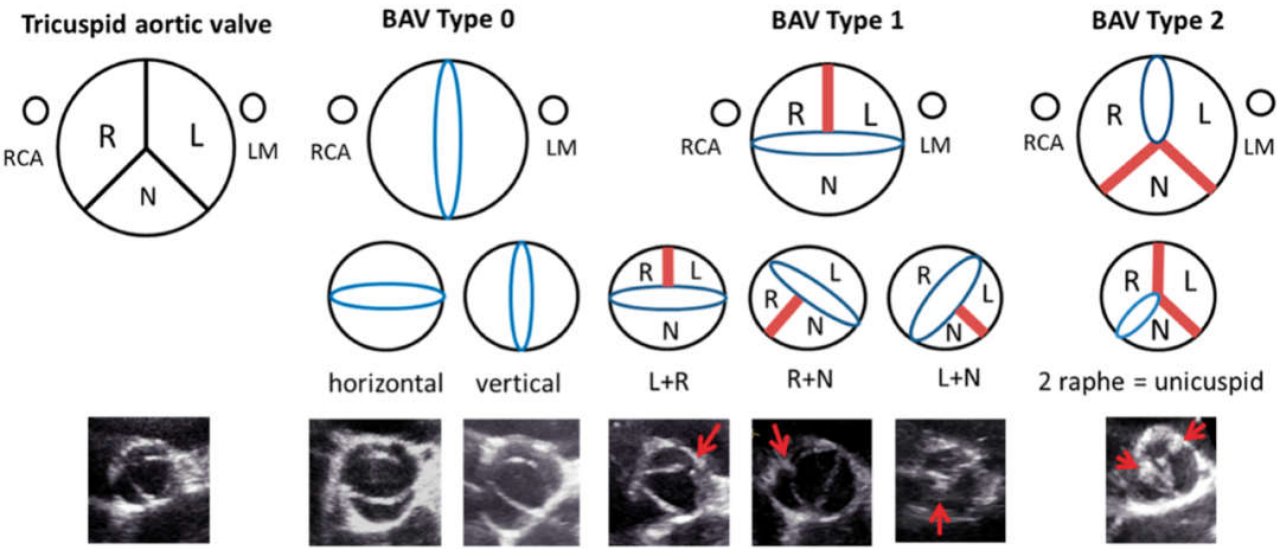


图5 附一张BAV分型图，方便对照

Table 2 Univariable and multivariable analysis of aortic measurements						
	Unadjusted analysis			Adjusted analysis ^a		
	Difference	(95% CI)	P-value	Difference	(95% CI)	P-value
Aortic annulus (mm)	0.98	0.78–1.18	<0.001	1.17	0.96–1.39	<0.001
Sinus of Valsalva (mm)	1.53	1.15–1.91	0.001	1.86	1.47–2.24	<0.001
Sinotubular junction (mm)	0.31	0.06–0.68	0.097	0.52	0.14–0.90	0.008
Ascending aorta (mm)	0.97	0.52–1.43	<0.001	1.05	0.57–1.52	<0.001

CI: confidence interval.
^aAdjusted for: age, gender, ethnicity, hypertension, dyslipidaemia, smoking, diabetes mellitus, and aortic regurgitation.

图6 该研究还同时发现，虽然同为BAV人群，但亚洲组的主动脉瓣环直径、SOV、STJ和AA明显更大

上文提到的BAV分类，最开始是德国研究者 Hans-Hinrich Sievers教授和 Claudia Schmidtke教授于2007年提出的，因此又称为Sievers分类，除了分类以外，其实文中还规范定义了很多专业术语，有兴趣地可以去读一下（就不会到现在还纠结于一些描述语言了

~~)。当然在影像学检查手段日新月异后，后续许多专家根据不同需求，譬如TAVR或手术的需要，更新了或进一步修饰了Sievers分类。



图7 Hans-Hinrich Sievers教授

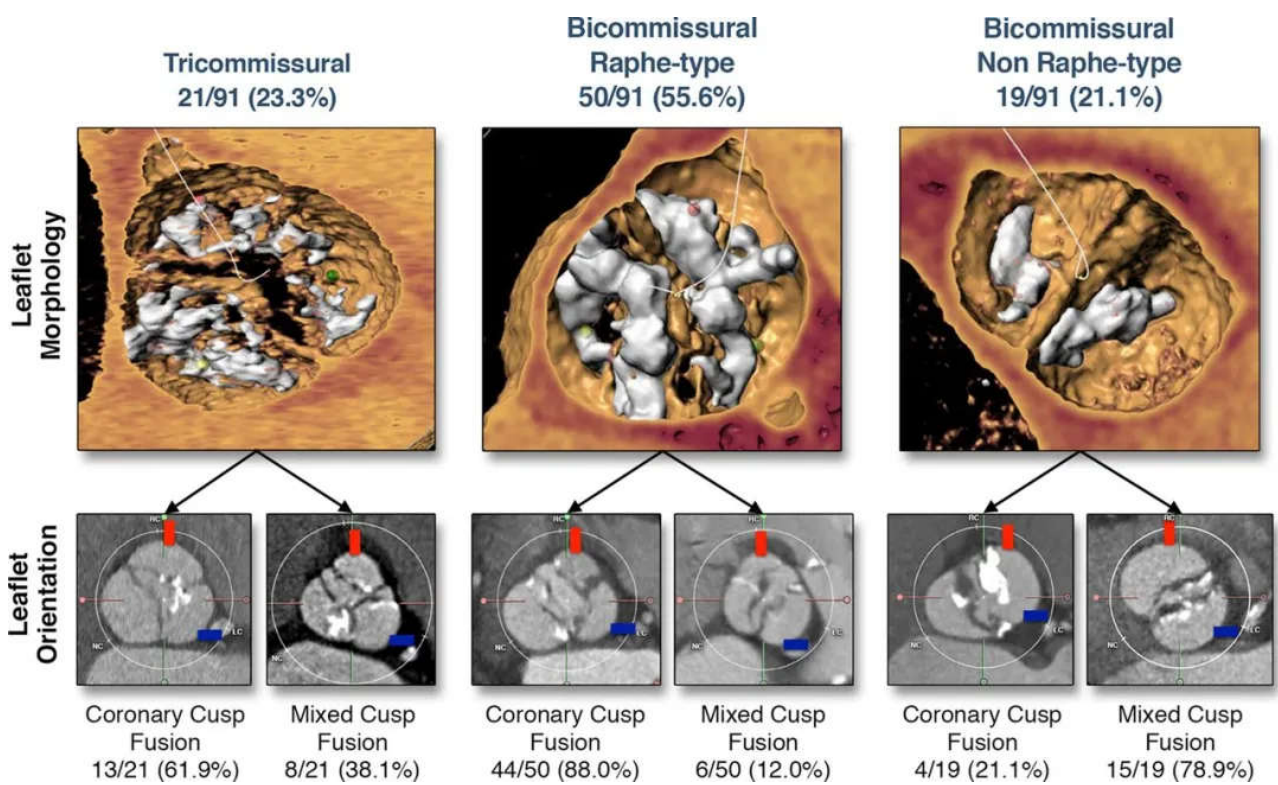


图8 ACCF 中外研究者联合制定的TAVR相关BAV分类标准

	MDCT		Schematic		Sievers et al.*	Jilaihawi et al.	Fusion Site
	systole	diastole	systole	diastole			
Bicuspid (n = 144)					Type 0 (n = 6)	Bicommissural, no raphe (n = 6/4.2%)	NA
					Type 1 (n = 138)	Bicommissural, complete raphe (n = 60/41.7%)	N-R 18.3% N-L 1.7% L-R 80.0%
						Tricommissural, incomplete raphe (n = 78/54.1%)	N-R 9.0% N-L 1.3% L-R 89.7%

图9 德国研究者Won-Keun Kim教授等根据MDCT对BAV的分类

主动脉置换固然可以简单粗暴地解决问题，不过对于年轻患者，再考虑到介入瓣膜不匹配、瓣周漏等一些并发症的风险其实并不低，更建议BAV修复术，毕竟天然的瓣膜结构还是最匹配的。BAV修复术作为外科置换AVR的主要替代治疗，已被多个研究证实可获得良好的效果。Mayo Clinic早期经验即证实，BAV患者外科修复后的5年生存率分别为96%和87%，其中，随访1、5和10年无须置换瓣膜者分别占96%、89%和49%，提示换瓣可以先等一等。在2017年 Cleveland Clinic Ahmad Masri教授的一篇系统回顾中，提示BAV患者外科治疗后的长期生存率，可达到正常无瓣膜疾病对照人群水准，也即治完可以和正常人一样活。（可以简单对比一下现在的TAVR治疗结果，不过一方面TAVR本身长期随访数据就少，更别提BAV这种细分领域；另一方面主要TAVR是高龄患者~~）

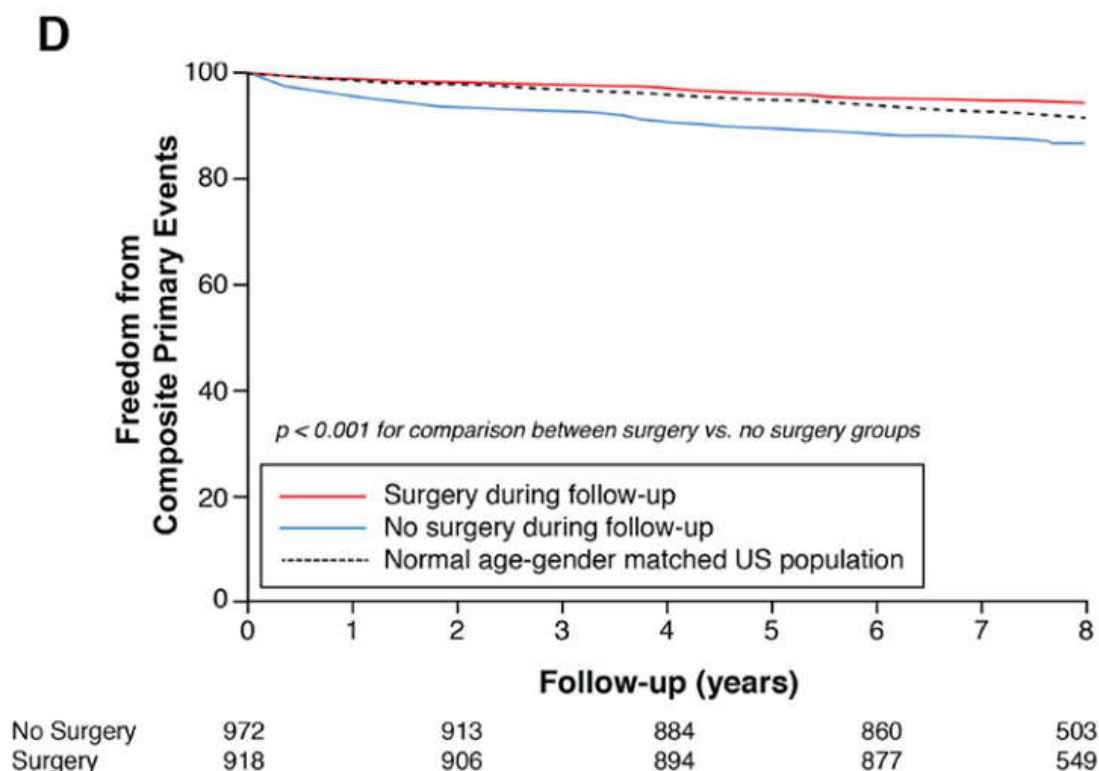


图10 Cleveland研究显示，BAV人群外科治疗后的长期生存率比较

不过，说一千道一万，患者碰到手术还是怕的，尤其心脏上开刀，笔者作为一个曾经动过刀子的患者，回忆麻醉醒来的时刻也是感慨颇多。作为患者，如何克服内心的恐惧，选择最适合自己的治疗方式，或者说如何制定最大收益的治疗决策，也是一门学问。今天姑且就聊到这。

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作者：肥肥黄馒头



微信号：CSTCVS1985

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