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LETTERS TO THE EDITOR

Sick Sinus Syndrome

Gillette's discussion of the sick sinus syndrome (1) is marred by several unsubstantiated statements and one flagrant misquotation. The tragic implications of needless pacemaker implantation in children make it imperative that the facts at issue be stated clearly and accurately.

1. The author states, "Instances of sick sinus syndrome in children who did not have congenital heart disease or cardiac surgery have been reported. At least one death in this group has been noted (2)." In fact, the case cited was that of a child born with transposition of the great arteries: the infant was cyanotic and seriously distressed at birth. A septostomy was performed, but the child died of aspiration of a nasogastric feed at the age of 1 week. How Gillette concluded that this child fell in the group "without congenital heart disease" is a mystery. The child suffered from intractable tachyarrhythmias during its week of life: no bradyarrhythmias were ever recorded. At postmortem there was evidence of hypoplasia and fibrosis of the sinus node: the authors speculated that absence of adequate sinus node function might possibly have played a role in the genesis of the tachyarrhythmias, but admitted this was only conjecture. In fact, there are no documented cases of death in children—or, for that matter, in adults—caused by the sick sinus syndrome, as such, in an otherwise healthy heart. We have conducted two computer-assisted reviews of the world medical literature and can find no exceptions to this observation. The excellent review of Shaw et al. (3) is worthy of study in this context.

2. Type I block is a normal response of the AV node to pacing; the rate at which such block appears varies widely from individual to individual and even from hour to hour in the same individual. It can usually be modified drastically or eliminated by atropine. *No data have ever been published correlating the rate at which type I block appears during atrial pacing with the subsequent development of AV node disease or dysfunction. In fact, no such data exist.* Josephson (4) noted that the various rates at which type I periods appeared in normal persons form a bell-shaped curve, beginning in the lower 70s peaking at about 130 to 140 beats/min, and falling off sharply at about 180 beats/min. He further points out that the wide range of paced cycle lengths at which type I block appears are a manifestation of differences in the basal state of the patients at the time of the study: he notes that occasional healthy young adults develop type I block at a relatively slow rate as a consequence of enhanced vagal tone. In a previous article, Beder et al. (5) claimed that "the atrial cycle pacing length at which AV nodal Wenckebach periods occurred was a particularly sensitive indicator of atrioventricular node abnormality," but cited no data to support this view: it would be more accurate to say that the specificity of this measurement is substantially nonexistent.

Because this misapprehension has emerged elsewhere it is worth expunging definitively at this time. Practically all normal individuals will manifest some type of AV node block in response to atrial pacing. The pacing rate at which the block appears and the form it takes will vary with a number of factors including the manner in which the pacing rate is increased, and the level of sympathetic discharge. It may be assumed that patients with overt AV node disease will manifest an exaggerated response to atrial pacing with block appearing at lower rates, or higher degrees of block. The overlap between the two subsets, however, is enormous, and has never been precisely studied for the very good reason that, for all reasonable clinical purposes, assessment of AV node function is accomplished by a knowledgeable inspection of the electrocardiogram recorded at various rates. Once overt AV node block has appeared, it can be characterized with remarkable precision by analysis of the surface electrocardiogram: the response of the AV node to atrial pacing adds no useful information. To tell a patient with normal AV conduction during adequate electrocardiographic study that future or impending AV node block could be predicted on the basis of the rate at which type I block was elicited during pacing would be to make a statement for which there is no foundation in scientific fact. The same is true of attempts to decide mode and site of pacing on the basis of this insignificant measurement.

3. Finally, Gillette makes the blanket statement that "In patients with syncope, if the heart rate decreases to less than 50 beats/min in an infant, 40 beats/min in a toddler or 30 beats/min in a teenager, it may be assumed that bradycardia is the cause of the symptoms." He bases these generalities on a series of eight cases, which means there could have been only two, or at most three, in each category (5). Two cases are an anecdote, not a statistic. In addition, with pacemaker insertion looming, it is hazardous to make such statements as these without qualifications and guidelines. Is the bradycardia transient or sustained? If the bradycardia is episodic, how long is it maintained? Does it appear only during sleep or does it persist in the face of physical activity? Have transient reversible causes of bradycardia been excluded before it is concluded that the condition warrants definitive treatment?

The almost miraculous evolution of pacemaker technology has brought in its wake a need for precise definition of the types of abnormality of impulse generation and conduction and their significance. The established criteria for valid scientific investigation will protect both the clinician and the patient from the consequences of overdiagnosis, unwarranted prognostication and meddlesome invasion: they should be followed scrupulously.

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5. Beder S, Gillette P, Garson A, Porter C, McNamara D. Symptomatic sick sinus in children as the only manifestation of cardiac abnormality or associated with unoperated congenital heart disease. Am J Cardiol 1983;51:1133–6.

Reply

I must disagree with Phibbs' statements in the strongest possible way. There are no flagrant misquotations in our article. We did make a mistake in Reference 1. We intended to refer to the work of Van der Hauwaert and Ector (1) rather than to the article by Fox et al. We must have inadvertently transposed references when preparing the manuscript.

Phibbs' computer-assisted reviews must be faulty because there are at least two other articles that describe children who died after having documented sick sinus syndrome. One is by James et al. (2) and the other is by Bharati et al. (3). The patient of Bharati et al. had a ventricular septal defect in infancy that closed spontaneously, and death did not occur until the child was 16 years of age. I have concluded that that patient did not have significant congenital heart disease anywhere near the time of death. These last two articles also give histopathologic correlation to the electrocardiographic and electrophysiologic findings and indicate that there is pathologic basis for sinus node dysfunction.

Phibbs does not seem to understand how we are using the response to atrial pacing. Probably everyone knows that it is normal for a patient to develop type I block during atrial pacing, and we certainly do. In pediatric patients who are sedated with Demerol and Phenergan before study, the pacing rate at which type I block occurs is higher. Normal values have been clearly developed and are published in our book (4). I agree that it is very difficult to predict future onset of atrioventricular (AV) block using this test; however, we have been very successful in predicting nononset of AV block using a rate greater than 120 beats/min. We feel that the AV node will continue to conduct normally for some length of time, probably at least 10 years, and we find it useful to insert an atrial pacemaker. If second degree block develops at a lesser rate while the patient is sedated with Demerol and Phenergan, we feel there is a possibility that AV block might develop at a later time and therefore we use a dual chamber pacemaker. We have not subjected this idea to random study because we feel that it might be dangerous to the patient to do so. We also feel that the difference between a dual and a single chamber pacemaker is not too great in cost or trauma to the patient and therefore prefer to use a dual chamber pacemaker if there is any question of AV conduction.

I think if Phibbs had thought his third point out carefully he would agree that the heart rates described are very conservative. These rates are intended for patients who are awake and they occurred for 6 seconds. When one is dealing with patients, one has to develop some criteria for action. I have developed these criteria over a long period of time working with pediatric patients. I have no concern that these rates are too high; I have some concern that they may be too low. When one is dealing with patients after the Mustard operation for example, in whom many centers have found a 5% or greater incidence of sudden death, it is important to establish criteria for pacemaker implantation to try to prevent such sudden death. I do not think that we can afford to wait until we have absolute statistical evidence that our criteria are exactly right before we can begin to use our common sense.

The main point of my article was that most causes of sudden death in children are now treatable and thus preventable. If we take Phibbs' point of view, we will not prevent many of these sudden deaths.

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4. Gillette PC, Garson A Jr. Pediatric Cardiac Dysrhythmias. New York: Grune & Stratton, 1981.

Correction

Table 1 on page 532 of the September issue of the Journal was printed incorrectly (Blaustein AS, Risser TA, Weiss JW, Parker JA, Holman BL, McFadden ER. Mechanisms of pulsus paradoxus during resistive respiratory loading and asthma. J Am Coll Cardiol 1986;8:529–36).

The following is the correct version of the table:

Table 1. Baseline Hemodynamic Variables

Case	Heart Rate (min^{-1})	Blood Pressure (mm Hg)	Pulsus Paradoxus (mm Hg)	Esophageal Pressure (mm Hg)		LVEF(%)		RVEF(%)	
				I	E	I	E	I	E
1	75	129/88	1	−4	0	73	72	41	50
2	70	96/57	4	−5	−1	71	63	39	36
3	80	128/81	7	−12	−4	69	73	54	37
4	61	117/68	6	−4	0	85	85	40	38
5	76	144/97	6	−5	−1	85	95	31	32
6	81	150/103	3	−3	+2	71	66	37	36
Mean	73.8	127/82	4.5	−5.5	−0.7	76	76	40.3	38.2
± SEM	3.3	8.5/7.6	1.0	1.5	0.9	3.2	4.9	3.4	2.8

E = expiration; EF = radionuclide ejection fraction; I = inspiration; LV = left ventricle; RV = right ventricle.