Novel life saving human circulation

To the Editor

Despite my current retirement from medical activities the experiences of what occurred in the Children's Hospital of Buenos Aires in the late sixties and early seventies are very clear in my mind.

It was a time in which the pursuit of excellence in the treatment of severe congenital heart defects prevailed.

At that time, Dr. Eduardo Galíndez, general pediatric surgeon, organized what would become the Cardiovascular Surgery Service. At the same time I was returning from the United States and Dr. William Kreutzer, a creative young surgeon who had been training with Dr. Cervini in San Pablo, was returning from Brazil.

Cardiology at the Children's Hospital had an excellent level. It had been a pioneer in Latin America, thanks to Dr. Rodolfo Kreutzer, who in 1936 created a Cardiology Service in the Children´s Hospital, something that was restricted to a handful of hospitals in the world. In this service there were prestigious clinical cardiologists specially Gustavo G. Berri and Eduardo Kreutzer.

In that atmosphere everything was enthusiasm with a creative working disposition far from any economic interest. So it was that in 1971 Willy Kreutzer took the risk of an unprecedented operation that collected the systemic venous return to the pulmonary circuit in a case of absent right ventricle due to tricuspid valve atresia. Just in case and considering the boldness of the procedure a small interatrial communication was performed that would be the first case of fenestration in this type of interventions.

Interestingly, and as it happened several times in medical discoveries (the case of our Eduardo Braun Menéndez and Page in Cleveland); Francis Fontan performed an operation theoretically similar to ours in France. I say theoretically because we took the lead not trying to ventriculize the right atrium by placing a valve at the inferior vena cava opening as Fontan did. We thought, after a few technical changes, that systemic circulation should have the lowest barrier to reach the lungs and this is how the atriopulmonary anastomosis originated. It consisted of a wide output opening between the right atrium and the pulmonary artery, free of obstructions and with normal resistances. It would all depend then on the left ventricle acting as a systemic pump in tricuspid atresia or as a single ventricle in further indications.

In his editorial Dr. Kreutzer (1) summarizes all there is to say about the long term results of this operation with its posterior amendments .To him I refer.

Dr. Alfredo Rodríguez Coronel

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To continue studies in patients with Wolff -Parkinson -White may erase the persisting lack of knowledge around the topic

To the Editor

I have carefully read the article by Stefani et al. (1) Risk assessments in patients with WPW are the basis of many studies mentioned in the discussion of this manuscript, and the results of this work are a contribution of important clinical value and for the interventional electrophysiologist who prefers the ablation of all WPW diagnosed. It is regrettable that they have not recorded the antegrade effective refractory period of the accessory pathway (AP).

I predict that the controversy related to the assessment results of patients with asymptomatic and symptomatic WPW and who should undergo radiofrequency ablation will be almost permanent; since the electrophysiological characteristics of AP are variable and unpredictable. Dorantes and colleagues (2) have suggested that the inability to induce arrhythmias in patients with AP does not guarantee that it will not be inducible in the future or that to provoke them necessarily implies their clinical appearance.

In a 4.2-year follow up period, Fazio et al (3), only refer 3.4% of patients with paroxysmal supraventricular tachycardia (PSVT) and Stefani et al (1) showed 11.9% PSVT in a 3.6 ± 3.9 year median follow-up. These same authors mention that 38 % of their sample presented with PSVT. When analyzing age, sex and location variables of the accessory pathway (1) no association with the occurrence of PSVT was found. These dissimilar results of PSVT presentation frequency with similar average follow-up time and the absence of PSVT relationship with age, sex and pathway location, show the unpredictability of accessory pathways. These electrophysiological properties of accessory pathways led Chávez González and Puerta, (4) after a review of pathophysiological conditions that recreate the increased accessory conduction pathways, to ask the following questions: What will the positive and negative predictive value of WPW asymptomatic patients, previously diagnosed as low-risk patients after electrophysiological studies, really be? How long

should they be followed-up to actually establish that they will not present a clinical arrhythmia?

Finally, both aspects will continue: ablation or not in asymptomatic and low risk WPW patients. There are arguments for and against ablation in asymptomatic patients. (5)

Pros:

- Silent tachycardia in apparently asymptomatic subjects (how long will it be silent?)
- Programmed electrical stimulation (PES) for risk stratification.
- Prophylactic ablation in high risk patients (fast in ducible orthodromic tachycardia, multiple path ways, short antegrade effective refractory period).

Cons:

- Severe asymptomatic arrhythmias are less fre quent than in Pappone's series.
- Electrocardiographic screening is costly (to find 165 children, 200000 subjects should be studied).
- Risk identification is difficult.
- In asymptomatic subjects, patient agreement for PES will not be achieved in many cases.

The study by Stefani et.al. (1) shows that we must continue to investigate the natural history of WPW, aiming to erase the lack of knowledge that may still persist around the topic.

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Authors' reply

We thank Dr. Chávez González for her observations on our work; it is always pleasant to discuss with colleagues the results of our investigations.

Regarding her remark " It is regrettable that they have not recorded the antegrade effective refractory period of the accessory pathway" it is worth mentioning that this study is a clinical follow-up that began a long time ago (36 years) when invasive electrophysiology was still in the most unpromising beginnings and its use in pediatrics definitely unforeseen. The other reason is that this study sought to determine clinical variables that could predict what kind of WPW can be left untreated during follow-up, and never raised the possibility of invasive examination as suggested by Pappone.

It is true that the inducibility of PSVT may be at random in each patient, but as electrophysiologists we must privilege the anatomical substrate conditions, and if an invasive study can establish that the patient is prone to PSVT it is because he has all the right conditions for an AV reentry, and whether he develops them or not is a question of time and of the whimsical physiology of each subject, but we must accept that he has the substrate to originate it. Knowledge of these predictive variables is useful although, as Dr. Chávez González mentions, it is met by an inherent limitation to human evolution. Accessory pathways are robust and vital with high conductivity when they initiate in children; then they become older, losing their properties and finally they disappear making it impossible to draw an extended prognosis without the physiological obsolescence that biology imposes upon each of us. (1)

There is a clear concept repeated in each series with extended follow-up of more than 10 years: accessory pathways pose a risk in patients with associated congenital or acquired heart disease and in those who are symptomatic for arrhythmias or syncope. (2) This is a very important finding, because it allows us to quickly identify the population at risk, regardless of the antegrade refractory period of the accessory pathway.

Published works historically yield different results regarding the association of sudden death with WPW. Some describe a follow-up mortality similar to that of the general population. (3, 4) Others, however, associate asymptomatic WPW with high mortality. (5, 6) Conducting routine invasive tests for risk assessment is a questionable exercise, as Wellens mentioned in his editorial on Pappone' work, and Dr. Chavez González also refers in his letter: 200000 subjects must be studied to identify 160 with "probable" risk. It has been observed that the risk factors that develop sudden death in adults are not clearly applicable to children. (7)

It should be noted that subsequently to the completion of our work, the consensus for the management of pediatric patients with asymptomatic WPW was published in 2012, considering non- invasive tests (ECG and stress test) for risk stratification. (8) There finally remains to accept the known limitations of noninvasive studies to evaluate the refractory period of a pathway.

According to the results of the most trained centers in ablation of accessory pathways where complications in large series are close to nil, it is debatable to submit a patient to an invasive electrophysiological study and not to ablate the accessory pathway thus ending the problem.

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