REVIEW ARTICLE

Evolutionary Process of the Fontan-Kreutzer Procedure

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SUMMARY

The evolutionary process that led to the Fontan-Kreutzer (FK) procedure was determined by the evolution of thought in pediatric cardiology since the pioneer Dr. Rodolfo O. Kreutzer created the cardiology department, with the support of Dr. Alberto Rodríguez Coronel in charge of catherization laboratory and anatomic pathologic department in the hands of Dr. Luis Becú.
The first atriopulmonary anastomosis (APA) was carried out in our enviroment without having knowledge of the Fontan work, who claimed the right atrium (RA) "ventricularization" by placing an inlet valve in the inferior vena cava (IVC). Our beginning was different, because it was based on the concept that the end-diastolic pressure of the main ventricle is the suction hemodynamics propellant source of this system.We never place a valve in the IVC and from the beginning we developed the concept of fenestration as atrial escape valve. In 1971 we describe two techniques of APA (one with homograft and the other with the patient's own pulmonary artery) and in 1978, a posterior direct APA as large as possible, without any valvular implant. Afterwards, from 1987, new and improved surgical techniques have been developed: the lateral tunnel (LT) and extracardiac conduit (EC), which have the advantage of being a conduit that does not cause loss of kinetic energy, as RA would do it, which is a volume chamber. Clearly, the FK procedure is the best option that we may offer today to the single ventricles (SVs), though certainly it is not easy to live with a single ventricle. In many patients, over time, it uses to present a progressive impairment of the system due to chronic low-output and the increase of central venous pressure (near the limit of the edema). However, this impairment does not occur in all cases, as it is shown by the longest-lived survivor of the world after 35 years with this system. Indeed, the FK procedure is a palliative surgery that determined one of the achievement in congenital cardiopathy surgery.

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Key words > Single ventricle - Fontan-Kreutzer

Abbreviations >

APA	Atriopulmonar Anastomosis	**IVC**	Inter-ventricular communication
RA	Right atrium	**FK**	Fontan-Kreutzer FK
TA	Tricuspid Atresia	**LT**	lateral tunnel
TPVBP	Total pulmonary ventricle *By-pass*	**IVC**	inferior vena cava
EC	Extracorporeal circulation	**SVC**	superior vena cava
ECC	Electrical Cardioversion	**SV**	Single ventricle
IAC	Inter-atrial communication		

BACKGROUND

On the 60th Meeting of the American Association for Thoracic Surgery (AATS) carried out in Boston in May 2009, at the resquest of the own AATS, I gave a presentation about the long-term evolution from patients operated with this technique after 40 years of life: "*The Fontan / Kreutzer procedure at 40: An operation for the correction of tricuspid atresia*". (1) Because in this historical account the Argentinian contribution was significant, the Director of the *Revista Argentina de Cardiología* suggested me to make it known locally.

Like any evolutionary process, comes up from a past that determines it. It imposes then, an account of the precedents that led to the Fontan-Kreutzer procedure (FK), that is to say, a total pulmonary ventricle By-pass (TPVBP).

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Argentinian pediatric cardiology had a beadle and an artifice named Rodolfo O. Kreutzer, Latin pioneer of the specialty, including European countries.

He Became a pediatrician at Hospital de Niños in 1921 (Figure 1) and then he became interested in rheumatic fever. In 1937 he created the rheumatic and heart diseases department (Figure 2) (fifth in pediatric hospitals in the world).

Until 1938, the year in which *ductus* surgery started, (3) congenital cardiopathies integrated the chapter on teratology, but that historical fact to identify opportunities for reparation and healing prompted a deep interest in the accurate diagnosis. Thus, in 1943, R. Kreutzer published five cases with venous angiocardiography (see Figure 2) and early fifties, the arterial angiography (Figure 3).

The development of surgical processes was progressive and fast with the treatment of "blue babies" with the Blalock-Taussig anastomosis (4) (1944) and later with the development of extracorporeal circulation (ECC) by Lillihei (5, 6), Gibbon (7) and Kirklin. (8)

The Cardiological Department at Hospital de Niños continued with its diagnostic development, deriving the surgical patients to Hospital Italiano and Hospital de Clìnicas, where Drs. J. A. Caprile and G. G. Berri, respectively, were responsible for the cardiological control (Figure 4).

In 1962 the "closed" surgery started, at Hospital de Niños and in 1963, surgery with ECC supported on solid bases, namely: a) cardiology, consisting of the aforementioned group, b) hemodynamics with Dr. González Parente and then with the scientific contribution of Dr. Alberto Rodríguez Coronel at his income after training in Chicago with Miller, c) pathology, with the invaluable contribution of Luis Becú after his internship at the Clìnica Mayo with Jesse Edwards, and d) surgery with Drs . Eduardo Galindez (Figure 5), Jorge Albertal (short term) and the undersigned, Guillermo Kreutzer, who returned to Hospital de Niños after having trained in San Pablo with the master of the Latin American Cardiovascular Surgery, Dr. E. J. Zerbini, who gave his full support, including a week of surgery at Hospital de Niños in which he participated together with his team.

The encouraging results and the development of new surgical techniques included the use of homografts (Figure 6) from the D. Ross works (9) for the treatment of pulmonary atresia with interventricular communication (IVC) and true arterial trunk. This required its preparation and sterilization by radiation at Centro Atòmico de Ezeiza, which demanded a huge effort, that he carried out personally. On the basis of the availability of a homograft, in July 1971, the new technique of atriopulmonar anastomosis (APA) came up.

PHYSIOPATHOLOGY OF THE FONTAN-KREUTZER PROCEDURE

As I reported, this type of surgery allows the survival of patients with single ventricle (SV). As it is logical to assume, living with a single ventricle is not free of problems, but it can certainly be said that life with this pathology is possible.

Wherefore this particular hemodynamic system works, are essential certain requirements that

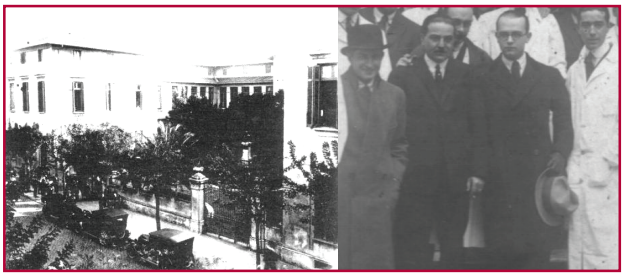

Fig. 1 Rodolfo O. Kreutzer at Hospital de Niños in 1921, when Gallo street had opposite direction.

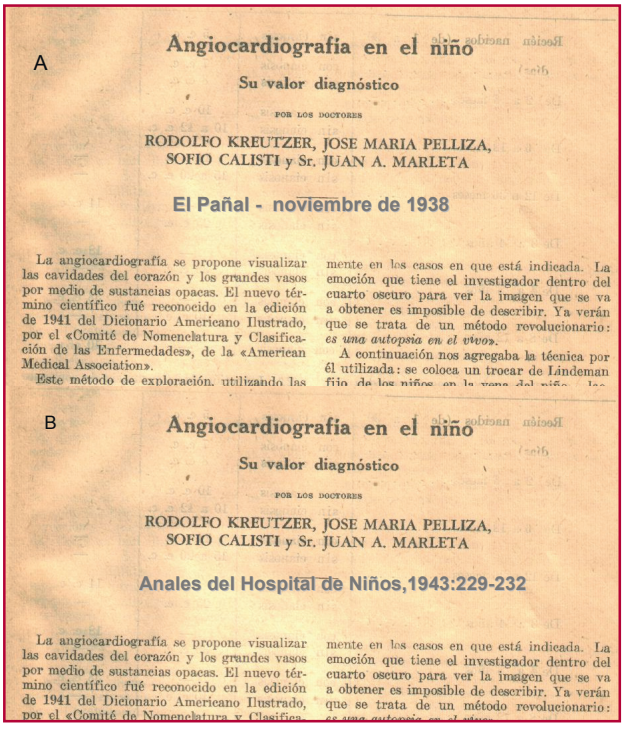

Fig. 2. Facsimiles of the magazine "El pañal" and "Anales del Hospital de Niños". A. Announcing the creation of Rheumatic and Cardiac Disease Department in 1938. B. Publication of the first angiocardiographies at Hospital de Niños in 1943.

Fig. 3. Rodolfo O. Kreutzer, in 1950, controlling by radioscopy the achievement of an aortography with contrast injection countercurrent by humeral artery in a patient with coarctation of the aorta.

Fig. 4 Cardiologic Department of Hospital de Niños in1964. From left to right: Drs. Juan A. Caprile, Ángel González Parente, Rodolfo O. Kreutzer, Gustavo G. Berri and Pathologist Luis Becú.

Fig. 5. Drs. Eduardo M. Galíndez and Guillermo Kreutzer. angiocardiographies at Hospital de Niños in 1943.

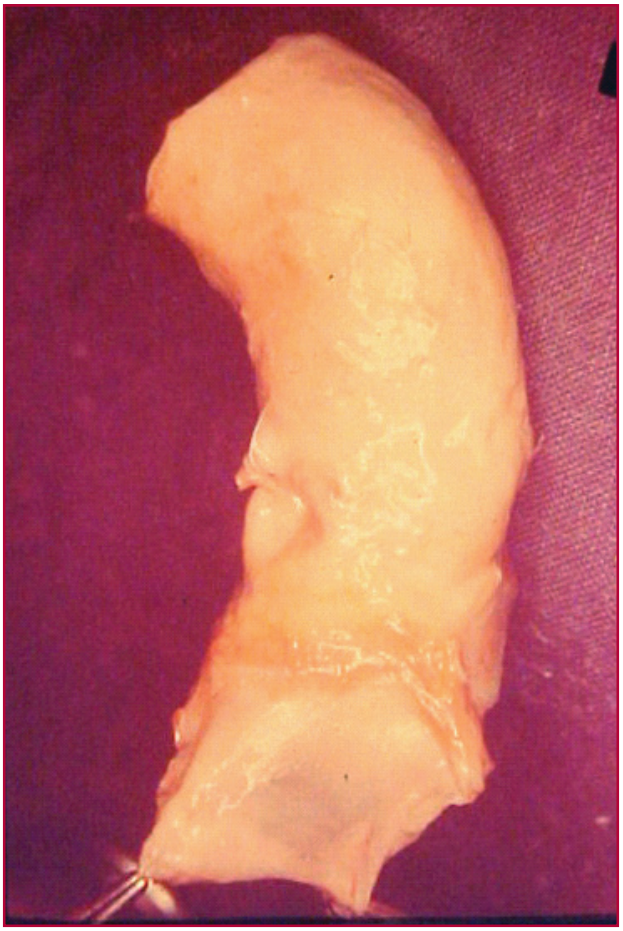

Fig. 6. Irradiated homografts used at Hospital de Niños since 1968.

allow venous blood to transpose the lung and reach the main ventricle without any disturbance. Unlike the biventricular heart, in this system the venous pressure exceeds left atrial pressure. The ideal is to maintain a gradient of about 6mm Hg, but with the levels of atrial pressure as small as possible. To achieve this, it is essential that the single ventricle is normal, as well as the systemic atrioventricular valve, the pressure and pulmonary resistances are low and surgical anastomosis mouths (the Glenn operation or the conduit) do not have any restrictions (Figure 7).

Another requirement is the presence of sinusal rhythm, as any type of arrhythmia causes an increased left atrial pressure disrupting the gradient and the system. Therefore, any alteration of one of these pre-requirements becomes dysfunctional to the system, which is manifested by signs of right heart failure with hepatomegaly, edema and, in some cases joint by protein-losing enteropathy.

HISTORY OF OUR FIRST ATRIOPULMONAR ANASTOMOSIS

Innovations usually occur when there is the need to solve a problem and in our case it was the attempt to solve oximetry and hemodynamics of univentricular heart with the development of the APA.

Our first APA was carried out without having knowledge of the Fontan works published six months before (January 1971) in *Thorax*, (10) magazine very widespread in our environment. In July of that year a three years patient came to our unit, dying, severely cyanotic with a tricuspid atresia (TA) with prior occluded Waterston's anastomosis (right aortopulmonary). Therefore, if he had thrombosed the right lung branch, it could not carry out left anastomosis. At that time it did not consider doing an anastomosis with ECC.

Opposed to Fontan, who to carry out his first procedure had two classic choices, systemic-pulmonary anastomosis or cavopulmonary anastomosis (Glenn), we only had two new possible operations: a) carry out an APA (11) or b) enlarge the interventricular communication. (12) We chose the first option and carried out a previous APA placing a homograft between the right atrial appendage and the pulmonary artery trunk (Figure 8A) without placing a valve in the inferior vena cava (IVC) or making Glenn operation, leaving a 6mm fenestration at interatrial *septum* level. Without doubt, it was the first fenestrated TPVBP worldwide. It was announced in the 5th Scientific Meeting of the Sociedad Argentina de Cardiologìa in August 1971 (11) and published in the *Journal of Thoracic and Cardiovascular Surgery*. (13) Simultaneously, Luis Becú pointed me that in the TA Ib, the pulmonary valve is usually normal according to his experience in pathologic anatomy. Accordingly, in December 1971, in another patient with TA Ib, lacking available homograft, we decided to carry out the APA with the patient's own valved

pulmonary ring, previously it was extracted from the right ventricle (Figure 8B), following the advice of Dr. D. Ross in order not to injure the first perforating septal in his surgery of aortic valve autograft.

With Rodríguez Coronel and Becú, our interexchange of views was focused on the following dilemma: "Does this procedure require a pump or a passage via?" (Figure 9). We reach to the conclution that RA lacks appropriate tissue to become a ventricular pump. We also consider the importance of the suction produced by the end-diastolic principal ventricle as a suction "bomb" of the pulmonary venous system.

During the commemoration of the centenary of the Hospital for Sick Children's in Toronto in 1975 I presented global experience TPVBP (35 cases) until then, (14) occasion in which I stated: a) "the partial or total closure of the oval foramen is subject to discussion. Its partial closure provides an exhaust valve to the right atrium, but it also causes certain degree of systemic desaturation, b) "the importance of sinusal rhythm reduces left atrial pressure, thus helping to obtain a reasonable gradient (6mmHg) between the right atrium and left atrium", c) "the left ventricle must be normal", d) "because in this system the venous flow is continuous, we doubt that the valves in the IVC and to the RA output work properly, increasing the peripheral venous pressure and the subsequent edema". This thought was opposed to Fontan's, who claimed the RA "ventricularization" putting valves at the inlet and outlet of the atrium. Rodriguez Colonel used to say that the pulmonary valve due to continuous venous flow, it would be never closed or opened completely, according to his words, it would be a *floppy valve*.

Finally, in 1978 I was convinced that the pulmonary valve was detrimental to this system, furthermore, it could determine some degree of obstruction. Accordingly, we totally change the surgical technique by introducing the direct posterior APA as wide as possible. After being sectioned the pulmonary trunk (without valve), it was passed behind the aorta, opened to the right pulmonary artery and carried out a laterally anastomosis to the opening made along the appendage on the roof of the RA. Previously, the IAC was closed and the tricuspid valve in the case of doble ventricular entry (see Figure 8C).

This technique was introduced in London in 1980, during the 1st World Congress of Pediatric Cardiology (15) and afterwards published in several international magazines. (16-18) One of the undisputable masters of cardiovascular surgery, J. W. Kirklin, sent us an affected letter with reference to this technique (Figure 10).

DEVELOPMENT OF NEW AND BEST SURGICAL TECHNIQUES

Several years later (1988), Marc de Leval (19) published the important concept about the loss of energy to reach both vena cavas at the RA (chamber of much larger volume). (20) Almost simultaneously with Castañeda,

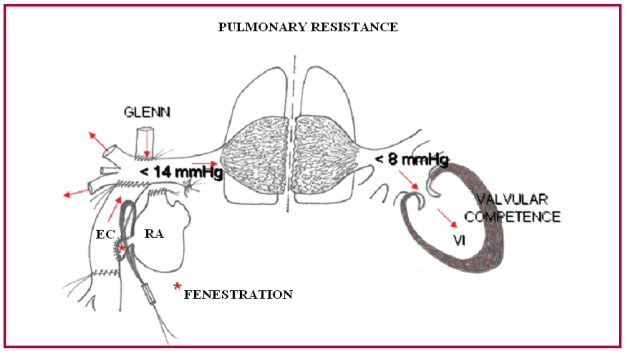

Fig. 7. Scheme of the proper functioning of TPVBP. The venous system (Glenn, EC and pulmonary artery) with venous pressure < 14 mmHg with gradient 6mmHg of difference with LA. In case if venous restriction at any level including pulmonary resistance, the system becomes dysfunctional, increasing venous pressure. The same happens if the pressure in the LA increases by ventricular dysfunction or AV failure or arrhythmia.

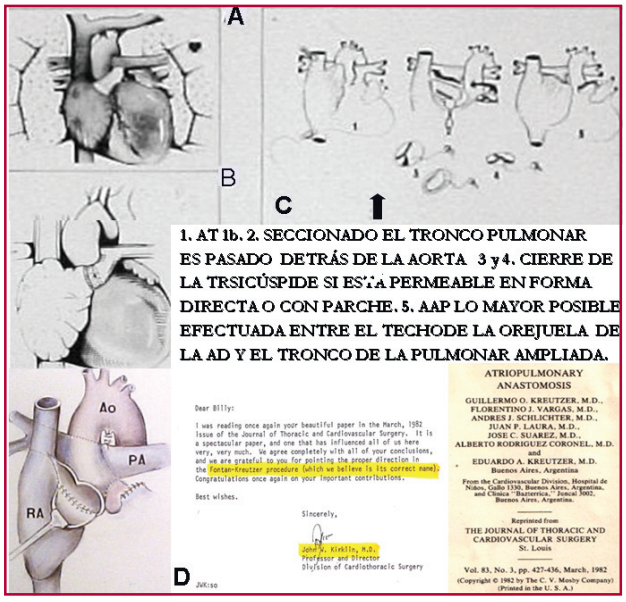

Fig. 8. Evolution of atriopulmonary anastomosis at Hospital de Niños. **A.** Prior APA with homograft (1971). **B.** Prior APA with the own pulmonary artery extracted from the outflow via of the right ventricle (1971). **C.** Later APA (1978). **D.** Conceptual setter sent by Dr. J. W. Kirklin referring to the later direct APA.

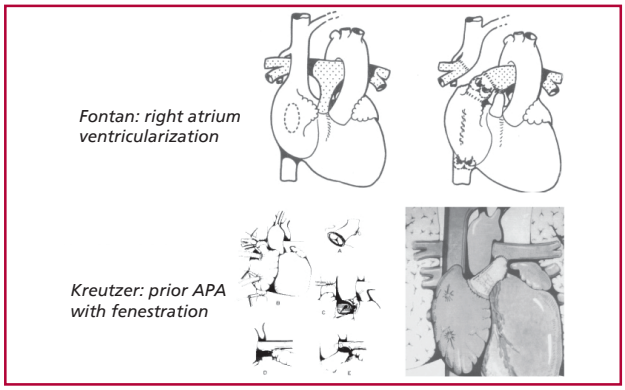

Fig. 9. During the seventies, the interchange of ideas at the Hospital de Niños was focused on the following dilemma: Do we need a pump or a venous transit via to the lungs ?

he spread the technique of lateral tunnel (LT) (21) (Figure 10A). Even though Puga had already used it a year before in an asplenia of single ventricle, (22) later, in 1990, Marceletti (23) popularized the technique of extracardiac conduit (EC) (Figure 10B) that Puga (24) had also used previously in 1988 (Figure 10 C).

Both techniques showed advantages over our APA, based on which it is a tube that runs the venous return and not a volume chamber; besides, prevents the complex atrial septation (Figure 10 D) which was necessary in the APA in the presence of a left stenosis or atresic valve (AV).

It is controversial which of the both techniques (EC or LT) is the best option. Both have excellent results (25) and therefore the choice depends on each surgeon's preference.

However, they should be considered some differences between the LT with Glenn operation and LT carried out after a hemi-Fontan surgery, (26-28) since the latter tends to produce more damage to the sinusal nodule area. The LT is used more in smaller patients and more frequently as the final stage after the Norwood procedure in hypoplastic left heart syndrome. EC has advantages for elderly patients taking into account the following: a)it does not need intra-atrial sutures, b) it is possible to carry out it without aortic clamping, c) the area of the sinusal nodule and *crista terminalis* (without suture line) remain at low pressure and, therefore, prevent injury and decrease the likelihood of arrhythmias, d) the possibility of closure of fenestration without the use of device following the technique proposed by our group (Figure 11) (29) and e) EC is a cylinder (18-20mm) with a similar diameter at its input and output. Therefore, it is different from the LT, which is a truncated cone at its base, the IVC is usually higher than its output by the SVC to the pulmonary artery. This is the reason why fenestration is usually needed and, when the hemodynamic status allows it, its posterior closuse requires device. (30) Moreover, EC is binding in the presence of bilateral SVC.

The possibility of enlarging the SVC with a pericardial patch creates risk of damage to the sinusal nodule artery due to its peculiar anatomy, because it may go ahead of the SVC, by behind or surround it. (31) On the other hand, the disadvantage of EC is that being a prosthetic tube cannot increase its diameter, which would be a problem for smaller patients, although TPVBP is usually indicated after eighteen months of life. Furthermore, it requires anticoagulant therapy.

Finally, the EC carried out after a Glenn operation, for prior pericardial adherences, it needs a higher level dissection of the IVC, which can lead to a phrenic paralysis, very detrimental to this particular system. The LT, with the use of total circulatory arrest, (28) without cannulation of the vena caves, it avoids this complication.

It has noted the advantage of eliminating the use of central venous lines to prevent thromboembolisms. (28) We favour the EC using the technique of "pericardial skirt" for fenestration, according to the proposed technique. (29) Before discharge the patient, if venous pressure allows it (<14mm Hg), it is possible to close it by adjusting the turnbuckle under local anesthesia, without the use of device (see Figure 11).

LONG-TERM EVOLUTION OF THE FONTAN-KREUTZER HEMODYNAMIC SYSTEM

The natural history of SV without surgery is truly disastrous (Figure 12 A).

Although it is not easy to live with a single ventricle, the FK procedure has allowed the survival, with improved prognosis and quality of life of these complex patients. (28) Even when they have chronic low-output, (32) sexual activity improves (which is a problem for cyanotic patients), it allows an almost normal life, but with limitations due to high venous pressure (12-14mmHg), near the border of the edema. Nevertheless, the palliative FK procedure represents the best surgical option that we may offer for this pathology.

However, it is clear that some adverse events, for multifactorial reasons, tend to occur after FK surgery, namely: right heart failure, arrhythmias that may require ablation and / or pacemakers, thromboembolism (33), protein-losing enteropathy , hypoalbuminemia (<3 mg /dl), (34) unexplained sudden death, (35) and so on. All these problems determine that, under the best circumstances, only 70% of patients are alive past 25 years and after overcoming several heart surgeries. (36)

In the presence of the system failure, echocardiography or, alternatively, cardiac catheterization determine the indication of reconversion, usually from APA to EC. Mavroudis experience (37) has systematized the reconversion, only it is indicated in symptomatic patients with multiple arrhythmic episodes, with no response to chronic treatment with amiodarone, or in the presence of thrombi in the RA or in the conduit.

Terminally ill patients, in renal failure, should be excluded from the possibility of reconversion. Heart transplant is not a better choice for the SV, but represents the last alternative for FK failed procedures. Its results in these patients are controversial. (38, 39) It has reported a progressive increase in pulmonary vascular resistance after FK surgery. (38)

In summary, remitting to the presentation at 60th Meeting of the AATS, in that time I had to indicate that for these patients it is difficult to exceed 40 years of life, which means being born before 1970, surviving to the initial palliative surgery and then to FK surgery. In our experience on 18 patients with FK surgery, born before 1970, only 7 are alive. Everyone had a SV with left predominance: six are TA and one left SV of double entry. In three of them it was successfully carried out the reconversion and now they have a normal life (Figure 12 B). One of these patients with

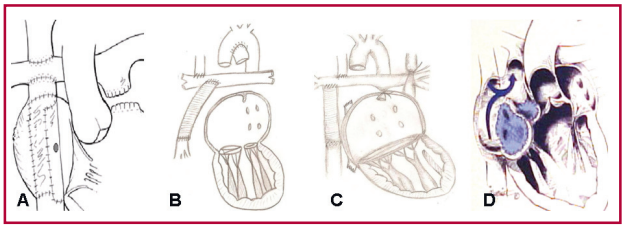

Fig. 10. A. Lateral tunnnel scheme proposed by de Leval and Castañeda. **B.** Extracardiac conduit scheme which was popularized by Marceletti. **C.** Extracardiac conduit previously employed by Puga. **D.** Scheme of complex atrial septation required in the APA in the presence of mital stenosis or atresia.

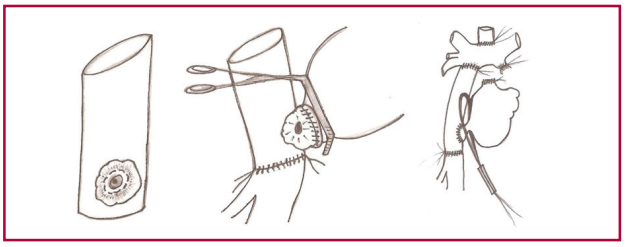

Fig. 11. Deferred occlusion technique of fenestration without the use of device.

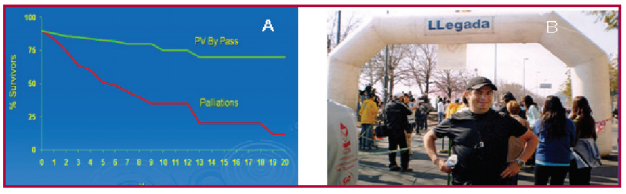

Fig. 12. A. Curve of postsurgical evolution of SVs. **B.** Pacient with TA Ib, happy after having finished a heel-and-toe race of 5Km to the year of reconversion.

previous FK surgery, operated in January 1975, today a lady of 53 years, is the longest-lived survivor of the world after 35 years of the palliative FK (Figure 13A). Her case was published (40) in November 2007 (Figure 13 B). For 20 years since her initial APA she had a normal life, she married, did not make pregnant and even divorced. After 20 years of surgery she began with arrhythmias and signs of calcification of the homograft. She was treated with amiodarone and anticoagulation, she rejected cardioversion indication until 2006, when she accepted it at the age of 49, being in terminal condition, severely symptomatic with hydrothorax (Figure 13 C), atrial fibrillation and large thrombus in the right atrium. Fortunately, ventricular function was preserved. Homograft and the thrombus in the RA were resected, besides carrying out a type Cox-Maze ablation. She was discharged 10 days after surgery (Figure 13 D).Nowadays, with 53 years, 4 years after the reconversion and the first 35 years of APA, she is asymptomatic, in sinusal rhythm and during ergometry she reaches 7 MET, an appropriate level considering that this is a person lacking any kind of training.

However, it is clear that the durability of FK procedure is limited. (41) Several hypotheses may explain the progressive impairment of this system at long-term follow-up, especially in complex SV that they are not TA, which have powerful left ventricles, and normal mitral valve.

Finally, they could summarize the adverse factors that lead to chronic increased venous pressure, namely:

a. **Inadequate surgical technique:** 1) classic Fontan (10) (Glenn + homovalve in the IVC and atrial output homograft between the appendage and the left pulmonary branch). 2) APA and its variants. (18) 3) the Bjork atrioventricular anastomosis. (12) 4) History of prolonged periods of systemic pulmonary ***shunts*** or PA banding.
b. **Progressive increase in pulmonary vascular resistance:** 1) AV valve pathology, particularly mitral atresia or stenosis with restrictive IAC. 2) Down Syndrome. 3) Distortion, absence or stenosis of pulmonary artery. 4) Living in height. 5) Chronic use of amiodarone. 6) Secondary lymphatic edema to high venous pressure. (32, 42) 7) Non chronic pulmonary pulsatility (43) and its effect on the distal pulmonary vascular bed. (41)
c. **Ventricular dysfunction (end-diastolic pressure> 12mmHg) due to:** 1) Prior volume overload. (32) 2) Inappropriate systemic ventricle: a) right (36) or indeterminate, b) cardiomyopathy, c) secondary myocardial fibrosis to prolonged periods of prior hypoxia, d) inadequate myocardial protection during previous surgeries. 3) Arrhythmias. 4) Atrioventricular valve insufficiency (more common in SV with heterotaxy or in hypoplastic left heart syndrome). 5) History of previous obstruction ventricular outflow tract or aortic arch. 6) Chronic low-output. (32)

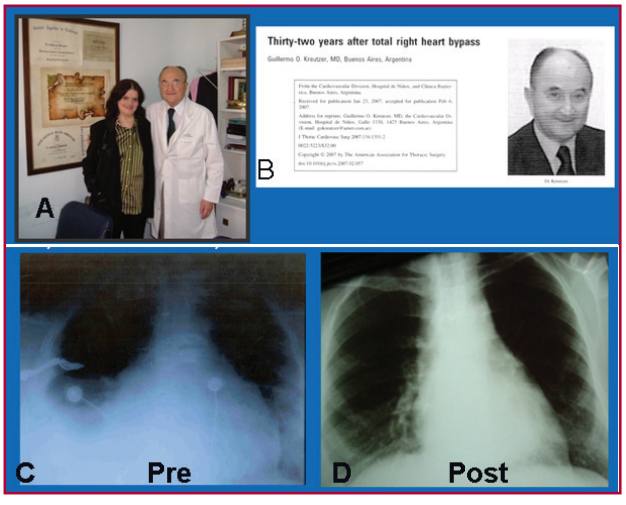

Fig. 13. A. Longest-lived patient of the World after 35 years of TPVBP and 4 years from reconversion. **B.** Publication on Journal of Thoracic and Cardiovascular Surgery. **C.** Prerreconversion radiography. **D.** Post-reconversion radiography.

CONCLUSIONS

With the improvement of surgical indications, new surgical and postoperative techniques, immediate and long-term results of FK procedure are excellent. (44) A total of 100 consecutive cases without mortality have been published. (28)

The future will be to improve furthermore surgical techniques, included the possibility of a partial mechanical device (45, 46) that heps to replace or supplement the pulmonary ventricle, or improve the feasibility and outcome of heart trasplant. Meanwhile, the radical palliative FK is the best option for SV despite its uncertain future. This palliative hemodynamics is definitely a huge success for congenital heart surgery.

RESUMEN

Sarcoidosis cardíaca: descripción de tres casos

El tratamiento invasivo de la fibrilación auricular en pacientes con miocardiopatía dilatada con sospecha de taquicardiomiopatía representa una decisión difícil y controversial. En esta presentación se describe el caso de un paciente de 57 años, internado por insuficiencia cardíaca congestiva progresiva. En el electrocardiograma se evidenció fibrilación auricular de alta respuesta ventricular y en el ecocardiograma, miocardiopatía dilatada con deterioro grave de la función del ventrículo izquierdo e insuficiencia mitral grave sin compromiso orgánico valvular. Se descartó enfermedad coronaria. Se planteó la ablación por radiofrecuencia como la mejor alternativa para su cuadro. El paciente recuperó ritmo sinusal, con el cual permanece desde hace 2 años, con evolución asintomática y mejoría de todos los parámetros ecoca rdiográficos.

Palabras clave > Fibrilación auricular - Cardiomiopatía dilatada - Ablación de radiofrecuencia por catéter

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