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The Paediatric Cardiology Hall of Fame

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There are many hallmarks of fame. In the medical literature, perhaps the most obvious, and certainly the most readily manifest, is to become eponymous. Thus, we constantly remember Wilhelm Ebstein, and Arthur Louis Etienne Fallot, when we describe patients with the malformations named in their honour. But these accolades did not accrue until many years after their deaths. Indeed, in the case of Ebstein, his now-famous description was but a minor case report amongst his overall bibliography of over 200 publications. It took almost a century before his name became widely used in the clinical context. Nowadays, however, one man's name is used even more frequently than those of Ebstein and Fallot. And he is very much alive. He is, of course, Francis Fontan (Fig. 1). Simply for the achievements which attracted the eponymous success, Francis would have deserved his induction into the Paediatric Cardiology Hall of Fame. But his activities in designing and pioneering his operation,^{1,2} in following its progress,^{3–5} establishing the criteria for its use,⁶ and establishing its optimal horizons,⁷ are just a small tranche of his overall achievements. These all round accomplishments extend far beyond the field of Paediatric Cardiology. Francis was, for 23 years, the Professor of Cardiovascular Surgery at the University of Bordeaux. Over this period, and during the apprenticeship leading to his appointment, his interests spanned all aspects of surgery on the heart and great vessels. His achievements in all these fields are impressive. For himself, however, he rates his greatest achievement as his part in the establishment of the European Association of Cardiothoracic Surgery, an organisation which is still growing in importance, and of

which he was the first President.⁸ It is for the operation, nonetheless, and for its modifications and ramifications, that he is, and will be, remembered amongst those involved with Cardiology in the Young. It was through our mutual interest in tricuspid atresia that I was fortunate enough, in 1974, to meet Francis. Since then, I have been honoured to count him as one of my mentors, and privileged to become one of his friends. As part of this friendship, I have enjoyed and shared his interests and expertise in wine, not unexpected for one based in Bordeaux, and similarly in golf. In this tribute, I will try and give some insight to all these multiple achievements.

Studentship and Apprenticeship

Francis was born in 1929, in the French burgh of Nay, in the foothills of the Pyrénées mountains. He

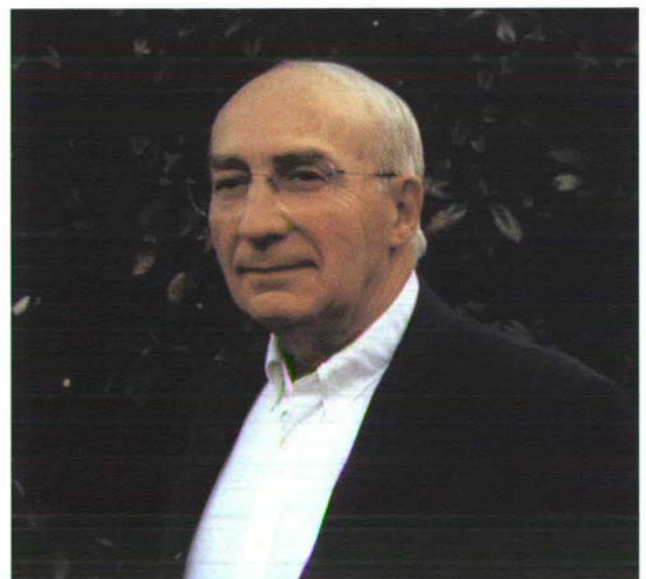


Figure 1.
Francis Fontan in London in June, 1999

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was born into reflected fame, since his father Victor was a noted professional cyclist. Victor Fontan achieved his greatest accolades as "King of the Mountains" in the Tour de France. On one occasion, he was leading the Tour itself by 15 minutes, but then suffered an accident. In those years, when support from the team and its mechanics was not as it is today, he was unable to recover. He was also a builder of bicycles, and a businessman, becoming involved in motor transport, so early childhood for Francis was comfortable. But then, when Francis was 10, France itself was engulfed in the conflict which was to become global. By this time, he had commenced as Boarder at the local Lycée in Nay. Initially, the Southern part of France was unoccupied by the German army. When it became occupied, he was able to continue his schooling, but no longer as a boarder, since his lodgings had been commandeered by the occupying troops. Despite the tradition of his father, he was not encouraged to cycle for sport, but became proficient at Rugby. His schooling was satisfactory despite the war, since soon after the *Année de la Victoire*, he was accepted at the age of 17 to enter the *Faculté des Médecins* at the University of Bordeaux. He spent six years as a medical student in Bordeaux, before continuing in the service of the University Hospitals as an *Interne des Hôpitaux*. Only 13 of one hundred students were accorded this honour. His internship occupied him for four years. He then entered the competition amongst interns to undertake a fifth year, and was sufficiently successful to be awarded the Gold Medal. Already he had selected his department for further studies and, in his third year of Internship, he entered the newly growing Department of Cardiac Surgery, working under his patron Georges Dubourg. Extracorporeal circulation as an adjunct to cardiac surgery was then an innovation. It had been introduced to France in 1955, and to Bordeaux in 1956. Surgery within the heart had been performed in Bordeaux since 1955, but supported by hypothermia rather than extracorporeal circulation. The use of the extracardiac circuit became the topic for his doctoral thesis. This was accepted, and circulated internationally, in 1959. Thereafter, progress was rapid. Already by 1959, he had become *Chef de Clinique* in the Medical Department of Cardiology, working under Professor Pierre Broustet. Then, in 1960, he became *Chef de Clinique* of the Surgical Department of Cardiac Diseases, returning to the service of Professor Dubourg. He became *Professeur Agrégé* in 1963 and, on the retirement of Dubourg in 1968, he assumed command of the

Surgical Department of Cardiac Diseases at the University of Bordeaux, based at the *Hôpital du Tondu*. It was here that he devised and performed the operations which have earned him his fame.

The Evolution of the Operation

Francis had become interested in the entity of tricuspid atresia following his experience with a young teenager who died in the Department of Medical Cardiology whilst he was an intern in Cardiac Surgery. Professor Broustet instructed Francis, together with his colleague Modeste Dallochio, to conduct the autopsy and harvest and describe the specimen.⁹ Georges Dubourg then suggested using this experience as the basis for a discourse on the currently available option for surgical treatment, which at that stage was exclusively palliative. The paper subsequently appeared.¹⁰ Within it, the opinion was expressed that, in the absence of a functioning right ventricle, the only corrective option at some future stage might become cardiac transplantation. The research needed to produce this paper made him aware of the cavopulmonary shunt procedure. He knew this anastomosis as the Bakoulev operation, since the concept of the shunt had been proposed in Moscow by Alexander Bakoulev, proved experimentally there by N. K. Galankin, and then performed clinically by E. Meshalkin. Already, however, Francis had come under the influence of Gerard Brom, Professor of Cardiothoracic Surgery at the University of Leiden in Holland. It was Gerard who drew Francis' attention to the earlier work of Glenn and Patino.¹¹ The concept of the cavopulmonary connection immediately triggered his imagination. He pondered that, if it was possible for the superior caval vein adequately to feed the right pulmonary artery, why should not the inferior caval vein be channelled to the remainder of the pulmonary trunk to supply the left lung? This concept was explored in the experimental surgical laboratory, using dogs and extracorporeal circulation. He closed the tricuspid valve, and by-passed the right ventricle. Apart from survival in the initial few hours, all operations proved unsuccessful. The results were never published. The work itself was performed between 1964 and 1966. The ideas then remained dormant. Then, by 1968, Francis had become Chief of the Department.

He was approached once more by Pierre Broustet, again concerning a young female adolescent with tricuspid atresia. The question was posed – what can be done? The initial options

offered were either the Blalock shunt or a Glenn procedure. But Francis then suggested the procedure tried previously, but unsuccessfully, in the dog laboratory. Despite the lack of experimental success, he was convinced it would work in the clinical setting. The patient had absence of the right atrioventricular connection, with concordant ventriculo-arterial connections, and naturally occurring pulmonary stenosis. Broustet, to his eternal credit, had sufficient confidence in the skills and judgement of his young surgical colleague to encourage him to proceed.

The operation was performed through a median sternotomy. At the same procedure, Francis fashioned an end-to-end anastomosis of the superior caval vein to the right pulmonary artery, closing the proximal stump of the caval vein. This, of course, was the Glenn shunt with which Francis was already very comfortable. He then proceeded by connecting the right atrium directly to the remaining part of the bifurcation of the pulmonary trunk (Fig. 2). Because of man's orthograde posture, an aortic homograft valve was inserted into the orifice of the inferior caval vein. Francis was equally familiar with the use of homograft aortic valves since, in 1965, he had established the first homograft bank on the Continent of Europe. The postoperative course was far from smooth, with pleural effusions and ascites as well as oliguria and, subsequently, anuria. Several periods of haemodialysis were required before the patient could be discharged, but this proved possible after several weeks of hospitalisation. She continues to do well, although conversion to an extracardiac Fontan procedure will likely be required in the near future. One further operation has been needed in the interim, namely removal of the stenotic and calcified aortic homograft valve.

Despite this success, no report was given of this initial experience. Then, in 1970, Francis was confronted by a second patient. This lady had survived to the age of 30 with no previous palliative procedures. She was very blue. This time the patient had tricuspid atresia with discordant ventriculo-arterial connections. The basic procedure was the same, but on this occasion, an aortic homograft was inserted between the right atrium and the pulmonary trunk (Fig.3). The postoperative course on this occasion was relatively smooth. She did well, became married, and gave birth to a healthy child. Unfortunately, after several years, she developed mitral incompetence. She died after a long and difficult replacement of the mitral valve.

The experience with these two cases was published in a French journal in 1971.¹ Between

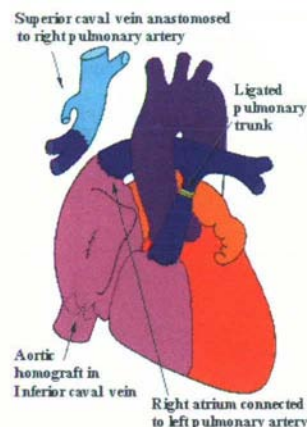


Figure 2.
The operative manoeuvres performed in the first Fontan procedure

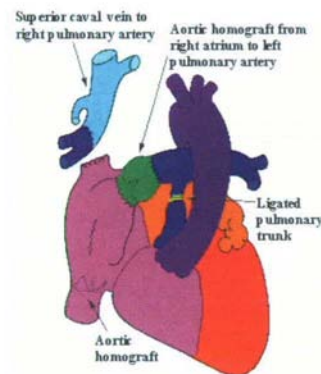


Figure 3.
The pathways constructed in the second operation, which included placement of an aortic homograft valve between the right atrium and the pulmonary arteries.

acceptance and publication, a third patient was seen, who underwent the same procedure but died. The autopsy revealed unsuspected pulmonary hypertension. On the basis of this experience, all three cases were published in *Thorax*, noting and emphasising the important contraindication to the surgical procedure.² It was the appearance of the report in the English language which then caught the imagination of the International Surgical Community.

Modifications of the procedure soon followed. Francis himself introduced the atrioventricular connection by means of a homograft conduit, but the procedure was modified surgically by Viking Bjork, avoiding the insertion of the conduit.¹² Guillermo Kreutzer offered the alternative of using the patient's own pulmonary valve so as to achieve the atriopulmonary connection.¹³ Francis had not initially used an outlet valve for the atriopulmonary connection, but had introduced this advance in his

second procedure. Of particular importance in the further development of the procedure in Bordeaux was the cooperation with Brom in Leiden. This had started in 1972 and, by the time of a Symposium held in London in 1977, forty patients had undergone the procedure. At this meeting, in addition to presenting the combined surgical results of Bordeaux and Leiden,¹⁴ Francis joined with his colleague Alain Choussat in promulgating the indications for the operation which soon became known as the "Ten Commandments".⁶ Already in the experience at Leiden, Francis had used the procedure in a patient with previously unsuspected double inlet left ventricle, and this option was popularised by Magdi Yacoub, who also emphasised the use of valves inserted into the orifices of the caval veins.¹⁵

Francis has followed further developments in "his" procedure with great personal interest. He is attracted by the concept of the total cavopulmonary connection, provided it is achieved by using the patient's own tissues to construct the venous channels, since he is concerned about inserting prosthetic material within the atriums. He is also concerned, however, with the potential danger of creating long suture lines, recognising the fact that such procedures unequivocally set the scene for subsequent arrhythmias. Because of this, he is particularly attracted by the concept of the extracardiac Fontan procedure, introduced initially by Carlo Marcelletti.¹⁶ Indeed, he collaborated with Marcelletti, Hanley, Mavroudis and others in preparing a joint presentation on this topic delivered at this year's meeting of the American Association of Thoracic Surgeons.¹⁷ Equally important in the evolution of his procedure was the analysis undertaken with John Kirklin at the University of Alabama in Birmingham. This culminated in the report of the "perfect" Fontan procedure.¹⁸ The activity continues even now. The next step will be an additional review of the next ten years of follow-up in the patients who formed the population for the "perfect" Fontan procedure. At the recent symposium held in London to celebrate the achievements of Christopher Lincoln, Francis reviewed again experience with "his" operation. In the course of his oration, he commented that, had he not thought of the procedure, then someone else certainly would, since the time was ripe. This is undoubtedly true. It is equally true, nonetheless, that he did think of, and successfully perform, the procedure. He comments that none of the patients undergoing his operation have been cured. This also is true. But very many patients throughout the world now owe their survival to his ingenuity and dedication.

Other Surgical Achievements

As Professor of Cardiac Surgery in the University of Bordeaux, it was essential that his interests span the entire field of Cardiovascular Surgery. His achievements over the field match the success of his operation. He was active in all types of congenital cardiac surgery, and achieved excellent results in procedures such as repair of tetralogy of Fallot or atrioventricular septal defect. He was skilled in atrial redirection procedures, and had no problem in switching to the arterial switch at the appropriate time. As already mentioned, very early in his career he recognised the advantages of the homograft valve. Introduced by Ross in the United Kingdom, and Barratt-Boyes in New Zealand, both pioneering the valvar substitute in 1964, Francis introduced the procedure to France in 1965. To gain the necessary experience, he visited Auckland, explaining in his lecture to the American Association of Thoracic Surgery¹⁸ that this reflected the advantage of being able to visit Tahiti en route. This far outstripped the attractions of the Channel Islands as the potential stop-off between Bordeaux and London. Encouraged by his experience, he assembled a large bank of homografts, establishing also the optimal means of harvesting the specimens.¹⁹ Between 1965 and 1986, his team harvested more than 1,100 valves, of which more than 500 were implanted, with about 400 being used in Bordeaux.^{20, 21} He also achieved excellent results in surgery to the thoracic aorta, with particularly important contributions related to traumatic rupture at the aortic isthmus^{22, 23} and aneurysm of the aortic arch.²⁴ He performed the first cardiac transplantation in France, and subsequently reestablished the cardiac transplantation service at the hospital which replaced Tondu at Haut-Leveque. In the most recent years, he focussed his interest on myocardial management and cardioplegia in adult cardiac surgery.^{25, 26} If he has any regret, it is in not spending one year in the United States during his period of training. Despite this, he was fortunate in those from whom he learnt, and who became his friends. Gerard Brom has already been mentioned, and was a huge influence. Equally important was the advice, encouragement, and subsequently the friendship, received from John Kirklin. Francis visited Kirklin for the first time whilst John was still at the Mayo Clinic. As described in his address to the American Association,¹⁸ after 1972 not a trip would be made to the United States without a visit being arranged to Birmingham. Happily, they continue. The overall topic of the presentation to the

American Association was "transplantation of knowledge". Francis has achieved great success also in this field. His first pupil was Eugène Baudet, still Professor of Cardiac Surgery at the University of Bordeaux. Eugène also followed in the steps of Francis to become the 12th President of the European Association of Cardiothoracic Surgeons. It is no mean achievement for Bordeaux to have provided two of the Presidents of this young but flourishing Association. During his tenure at Bordeaux, many other young surgeons and physicians flocked to learn from Francis. I was fortunate to visit on several occasions, often with Michael Tynan, and the advice and friendship offered was considerable. One particular aspect sticks in my mind, which I subsequently found was handed down from one of his own mentors. When you think you have finished a manuscript, put it away for four to six weeks, and then re-examine the work. Any deficiencies will then be much easier to appreciate. Jan Quaegebeur also acknowledges his particular debt for experience gained in Bordeaux, and during visits made by Francis to Leiden. Perhaps the biggest accolade was when a group of his young pupils persuaded him to leave the University of Bordeaux in 1991 and join them at the relatively new practice created at Clinique St Augustin. This happy association also continues to flourish.

Administrative Achievements

As mentioned already, Francis himself considers his greatest achievement to be the part he played in the establishment of the European Association of Cardiothoracic Surgery. He described some of the steps involved in the address he gave as President at the first Annual Meeting.⁸ The success of the Association reflects his own involvement, and he is particularly gratified by the establishment in his name of the Francis Fontan prize. Now awarded annually, the successful applicant is permitted to spend one year of training as set out in the application to the Prize Committee. Perhaps equally important for Bordeaux was his part in the establishment of the Hôpital Cardiologique at Haut-Levéque. This hospital, newly created in 1978, replaced the old Hôpital Tondu. It was built over a period of two years, following long and arduous discussions and deliberations with local, University, and national politicians, all requiring the very best of his considerable diplomatic skills.

The Inner Man

Recognition has deservedly come to Francis, largely in terms of the acceptance by the International community of his considerable achievements. In this respect, in addition to his inaugural Presidency of the European Association, he is proud of his Honorary Professorship at the University of Alabama in Birmingham, his Honorary Fellowship of the American College of Cardiology, when his marshall was John Kirklin, his Honorary Membership of the American Association for Thoracic Surgeons, unusually awarded ten years before he was invited to give the guest address,¹⁸ and his Honorary Membership of the Society of Thoracic Surgeons of Great Britain and Ireland. He is equally proud of his family. He described in his guest address the importance of the encouragement he received from his own father, and how he tried to pass this on to his children. Now grown, they are successfully launched on their own careers in law, business, and the wine industry, respectively. He is now additionally a contented grandfather, with four grandchildren already thriving. He remains remarkably active, and retains a thriving surgical practice at St Augustin. Concentrating largely now on coronary arterial surgery and valvar replacement, he continues to hold his own in terms of results and outcomes with his young colleagues. To me, he seems to have changed little over the years of our friendship (compare Figs 4 & 5). He is as active now as he ever was, albeit that his surgical schedule now leaves him more time to indulge his hobbies. With his wife, Maryse (Fig. 6), he spends enjoyable and competitive hours on the golf course, and together they enjoy travelling. His newest venture, nonetheless, together with his son Edouard, his wife Maryse and one of his friends is the reconstruction and development of Chateau l'Ermitage. Derelict until recently, Francis has revitalised this vineyard in Sauternes. Now producing wines of the finest quality (Fig. 7), his status as co-owner of the Chateau permitted him to taste on equal terms with the Marquis de Lur-Salaces, until recently the owner of Chateau Yquem. If l'Ermitage has yet to match Yquem in the vinous Hall of Fame, there can be no doubt but that Francis Fontan is Premier Cru Classé in the Paediatric Cardiology version. It is my hope that he long continues to enjoy his semi-retirement, and his full retirement should he finally decide to take it. I look forward to many more rounds on the golf course, and to drink with him even more bottles of the finest Bordeaux wine!



Figure 4.
Francis photographed with myself during a visit to Brompton Hospital in March, 1988.



Figure 5.
Francis and myself photographed in June, 1999



Figure 6.
Francis and his wife, Maryse, photographed in London in June, 1999



Figure 7.
A bottle of the 1992 vintage of Chateau L'Ermitage. This wine was ranked first of the Sauternes 1992 in the Gault and Millau magazine of September 1994, and obtained the Prize of Excellence at the Vinalies International in March, 1995.

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