Atrial Switch Operation: Past, Present, and Future

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The atrial switch operation was developed by the efforts of many surgeons, with the most notable contributions made by Blalock, Hanlon, Albert, Baffes, Senning, and Mustard. The atrial switch operation was the first definitive repair for patients with transposition of great arteries and produced good results. Although it is rarely performed today, the atrial switch is not merely of historical interest as there remain a few important indications for this operation. A thorough understanding of the atrial switch is still required for surgeons dealing with complex congenital cardiac malformations. Herein we summarize the history, review long-term results, and discuss the future of the atrial switch operation.

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These heroic men whose life work marked epochs in medicine we think of as individuals, but what they accomplished singly was perhaps of less importance than the inspiration they gave to the group of men who followed them.

—William James Mayo (1861–1939) [1].

Early surgical attempts to palliate transposition of the great arteries began before the advent of the open heart surgery. Many surgeons contributed to the development of the atrial switch operation. The most prominent and notable contributions were made by Alfred Blalock, Rollins Hanlon, Harold Albert, Thomas Baffes, Ake Senning, and William Mustard. Their ideas evolved into the operation that provided hope to many parents and saved the lives of many children. Today the atrial switch is largely replaced by the arterial switch operation. Consequently, surgeons may give little thought to the work of many individuals whose efforts culminated in a very successful surgical procedure that completely changed the outcomes for children with transposition. However, it is these practical dreamers who contributed to the progress of our specialty and, above all, served children who would not have survived without surgery. Herein, we describe the chain of procedures that wended its way through history and discuss the present and future of the atrial switch operation.

MISCELLANEOUS

Past

Atrial Septectomy

In the late 1940s Alfred Blalock, working with C. Rollins Hanlon at Johns Hopkins (Fig 1, A and B), anastomosed the pulmonary veins to the superior vena cava (SVC) and to the right atrium in dogs [2]. Blalock considered that this type of anastomosis might be feasible for patients. The animal experiments were performed with the strong technical support of Vivien Thomas, Dr Blalock's longtime laboratory associate [3, 4]. During those early experiments Blalock, Hanlon, and Thomas conceived the operation of redirecting venous blood flow inside the atria while leaving the transposed great arteries alone. In 1950, Blalock and Hanlon published the first clinical experience with their procedure that became known as the Blalock-Hanlon operation [5].

The Blalock-Hanlon operation (Figs 2 and 3) allowed intraatrial mixing of the pulmonary and systemic circulations by creating an atrial septal defect. It was the first palliative procedure that permitted survival of children with transposition of the great arteries [5]. In older infants, the Blalock-Hanlon operation was well tolerated and the mortality rate was relatively low. However, it was associated with a high mortality in the first few weeks of life [6]. Doctor W. Sterling Edwards from Birmingham, Alabama, modified the Blalock-Hanlon operation to provide obligatory intraatrial mixing by suturing the surgically mobilized flap of atrial septum to the left side of the right pulmonary veins [7]. In 1965, Edwards reported this operation performed in 18 infants (Figs 3 and 4). The Edwards modification of the Blalock-Hanlon operation was described after the Senning and Mustard operations were already in use. Although the Blalock-Hanlon operation and its modifications were eventually superseded by Rashkind's technique of enlarging the foramen ovale with a balloon catheter, they produced good intermediate-term palliation and were applied in some institutions for decades after their first description [8, 9].

Early Arterial Switch Attempts and Techniques of Venous Return Transfer

The initial attempts to perform the arterial switch operation were universally fatal [10, 11–14]. On January 17, 1952, Mustard operated on a 3-month-old girl with transposition, using a monkey lung as a biological oxygenator, and attempted an arterial switch [11]. Mustard per-

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Fig 1. *(A) Alfred Blalock (1866–1965); (B) C. Rollins Hanlon (1915—). (Courtesy of Dr C. Rollins Hanlon.)*

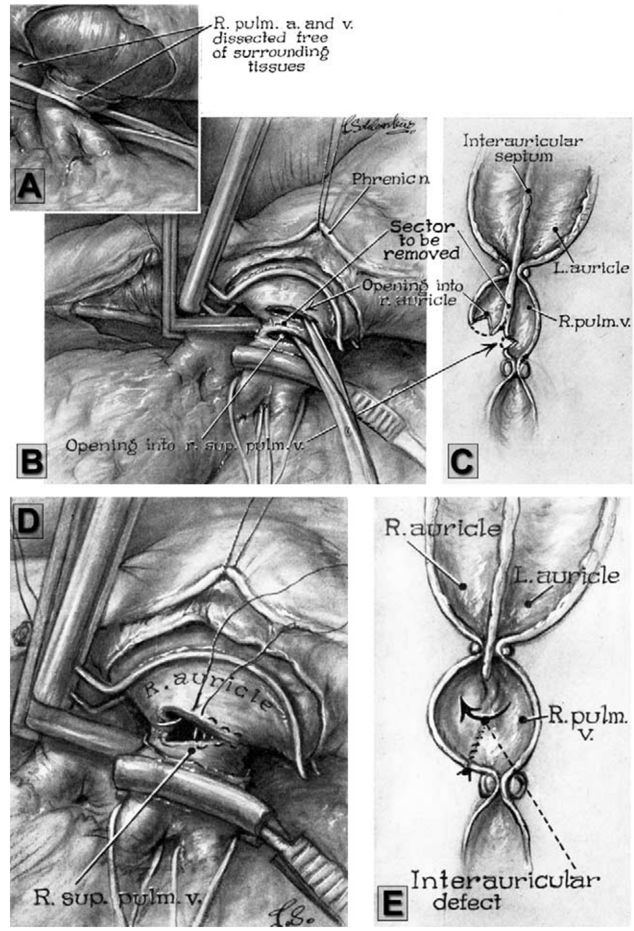

Fig 2. Blalock-Hanlon operation. With the right pulmonary artery and superior pulmonary vein dissected free from the surrounding tissue (A), right atrium, pulmonary veins, and pulmonary artery are clamped (B). Parallel incisions are made in the right auricle and in the anterior wall of the right pulmonary veins. These two incisions expose the edge of atrial septum and allow for its excision (B and C). The edge of the atrial septum has been excised and the incision in the right atrium and pulmonary veins is closed with sutures (D and E). (L. auricle = *left auricle;* R. auricle = *right auricle;* R. pulm. a. and v. = *right pulmonary artery and vein;* R. sup. pulm. v. = *right superior pulmonary vein.) (Reprinted from Blalock A, et al, Surg Gynecol Obstet; 1950;90:1–15 [5], with permission.)*

formed a transfer of the left coronary artery to the neoaorta, leaving the right coronary artery intact and arising from the neopulmonary artery. He thought that pulmonary arterial pressure would be sufficient to provide coronary flow for the right ventricle that would be under low pressure supporting the pulmonary circulation after the arterial switch. By November of the same year, Mustard had performed a similar operation on 7 babies. None of the infants lived for more than a few hours. On November 10, 1952, Bailey attempted an arterial switch in a 7-month-old boy, who also died postoperatively [12]. In 1955, Senning attempted an arterial switch procedure with no success [10]. The obstacle to anatomical correction of transposition was the difficulty in transferring the origin of the coronary arteries, and most surgeons focused their effort on switching the venous return. In March 1952, Walton Lillehei anastomosed the right pulmonary veins to the right atrium and the inferior vena cava to the left atrium [15] in 4 babies, and 2 survived. In 1955, he reported a series of 32 children, 7 of whom survived for 15 to 36 months after surgery.

In 1956, Thomas G. Baffes, working with Willis J. Potts in Chicago, described his technique [16]. Baffes used an aortic homograft to join the IVC to the divided proximal end of the right pulmonary veins and anastomosed the distal end of the divided right pulmonary veins to an incision in the right atrium (Fig 5). Baffes performed his first successful clinical operation on May 6, 1955. Within a year, Baffes operated on 38 patients with 19 survivors [17]. All of these operations were performed without cardiopulmonary bypass. Baffes intended to perform a second stage that would switch the SVC with the left pulmonary veins, but this was never performed clinically.

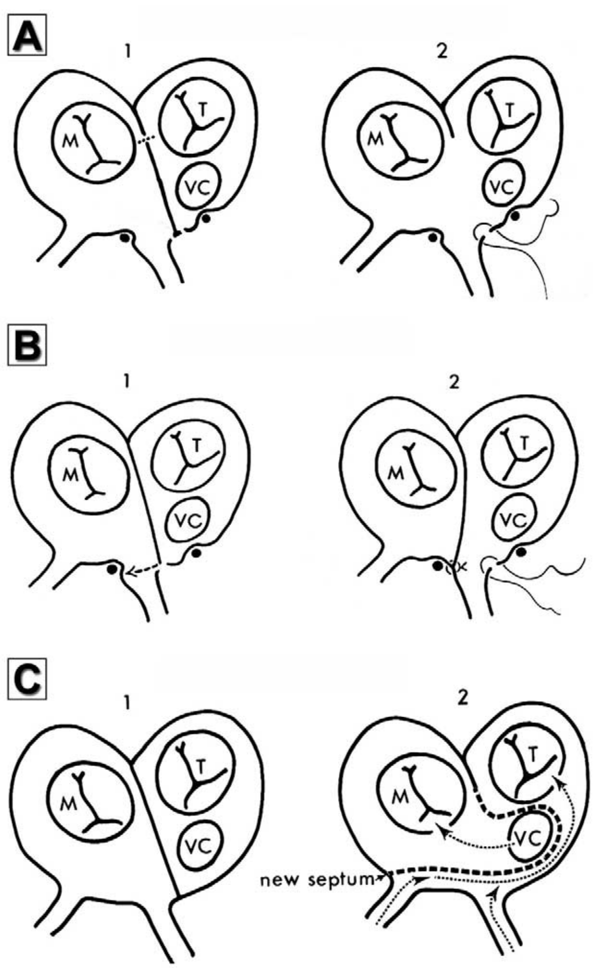

Fig 3. Schematic diagrams of (A) Blalock-Hanlon, (B) Edwards, and (C) Mustard operations. Atrial septum is excised (A and C) or shifted (B1) and sutured (B2) to the posterior wall of the left atrium. The thick black dots *indicate the position of the clamp. The* arrow *in (B) indicates the direction of the translocation of the atrial septum. The* arrows *in (C) represent blood flow.* 1 = *diagram showing atrial septal position;* 2 = *diagram after completion of each operation.* (M = *mitral valve;* T = *tricuspid valve;* VC = *vena cava).*

Baffes' operation remained an excellent treatment option for transposition over the next 10 years.

The idea of physiologic correction of transposition using an intraatrial baffle came from a brief paper published in 1954 by Dr Harold M. Albert [18]. Albert used a flap of the atrial septum for systemic and pulmonary venous flow redirection in dogs (Fig 6). Of 20 dogs operated upon with hypothermia and circulatory arrest, 15 survived from 22 minutes to 4 weeks before dying or being sacrificed. He stated that 2 animals survived for 2 and 4 weeks without significant stenosis or thrombi, but with effective transposition of the venous return. Although Albert used atrial septal tissue in all his experiments, he made a reference to an alternative technique (Fig 6, E) using a "plastic prosthesis" in his original report [18]. However, using a plastic prosthesis seemed unnecessary in the presence of autologous pericardium. When asked to comment on use of an artificial material for atrial baffle, Mustard replied jokingly: "Use pericardium! The only excuse for using Dacron is if you drop the pericardium on the floor" (Mustard to Van Praagh, personal communication). Albert did not attempt his operation clinically. Yet Albert's principle became the foundation for the atrial switch operation. Subsequently, Mustard always credited the idea for his operation partly to Dr Albert [19]. Inspired by the experimental work of Albert, Oscar Creech and colleagues from Tulane University in New Orleans attempted an atrial switch in a 9 year-old boy on December 4, 1956 (Fig 7). They used a polyvinyl (Ivalon) baffle and extracorporeal circulation to transfer blood flow from the pulmonary veins into the tricuspid valve [20]. The patient achieved 100% saturation and regained consciousness. Unfortunately, the child had a cardiac arrest in the recovery room 12 hours after surgery and died. In 1966, after acknowledging the work of his predecessors, Mustard stated: "...in my manuscript I gave credit to Dr Albert for his original principles; the only problem with these procedures was that they did not work" [21]. The similarities between the Albert and the Mustard procedures were striking indeed. Mustard once was heard saying: "After all, the only difference between my operation and the Albert procedure is that mine works" (Mustard to Van Praagh, personal communication).

It was apparently Dr Potts who provided the guidance and support in the initial research performed by both Baffes and Albert in the laboratory of the Children's Memorial Hospital in Chicago [22]. Baffes first met Potts in 1952 and worked with him first as a resident and then as an associate. Later, Dr Baffes wrote: "When I first broached to Dr. Potts the idea of designing a partial venous correction for transposition of the great arteries, he puckered up his mustache, as he was wont to do, and lapsed into deep thought. After a while he said: 'Transposition represents about half of the defects in cyanotic children coming into this hospital. We haven't been able to do much for any of them, except those special ones having pulmonary stenosis with ventricular septal defects.' He thought a little longer; then his eyes twinkled with excitement. 'Go to it! I'll get the necessary funds.' Thus I began many happy years of inquiry and discovery under the guidance of the master surgeon" [22]. Baffes extensive work and clinical success demonstrated the feasibility of, at least, partial extra-atrial switch of the venous return.

On March 20, 1957, Alvin Merendino, at the University of Washington in Seattle, first attempted to clinically apply a modified Albert's technique. Instead of a bilobed flap of the atrial septum as described by Albert, Merendino used a premolded atrial septal prosthesis. With use of cardiopulmonary bypass and elective cardiac arrest, Merendino excised the atrial septum and inserted a plastic baffle to switch the venous flow in a 6-year-old girl. The girl died owing to irreversible ventricular fibrillation after the prolonged operation [23]. A second patient died of cerebral embolism. Nonetheless, Meredino's operation fueled the interest in the atrial switch operation.

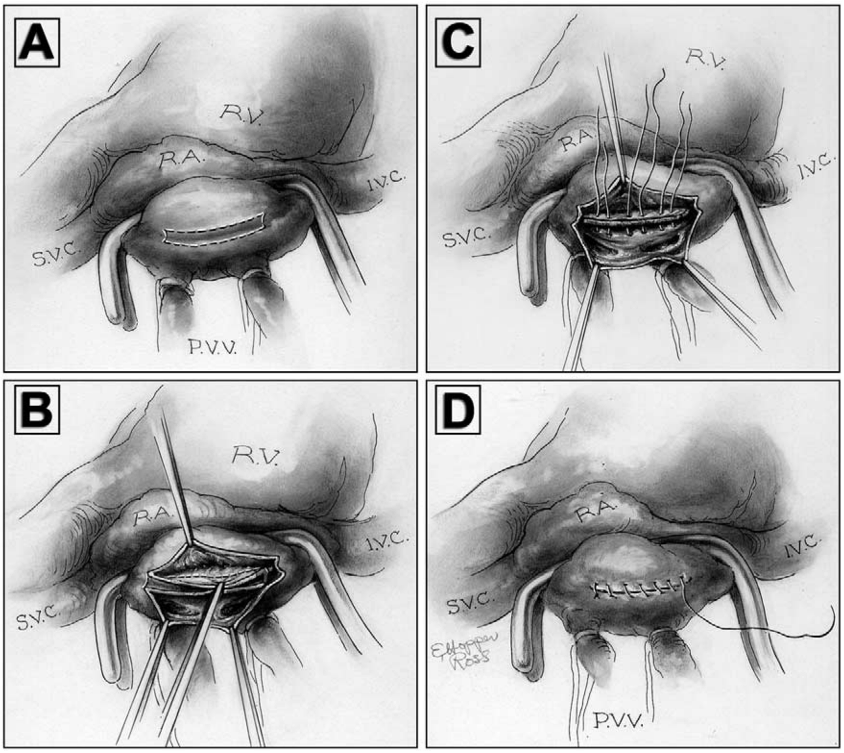

*Fig 4. Edwards operation. (*A*) Adjacent portions of the right and left atria are clamped. Heavy silk loops occlude the right pulmonary veins and a tourniquet obstructs the right pulmonary artery (not shown). (*B*) The right and left atria are incised on either side of the interatrial septum, extending above and below the right pulmonary veins. The interatrial septum is dissected and trimmed sufficiently to produce a mobile flap. (*C*) The septum is sutured to the wall of the left atrium behind the right pulmonary vein orifices. (*D*) The atrial incision is closed and circulation restored. (*I.V.C. *= inferior vena cava;* P.V.V. *= pulmonary veins;* R.A. *= right atrium;* R.V. *= right ventricle;* S.V.C. *= superior vena cava.) (Surgical palliation in complete transposition of the great vessels. Experience with the Edwards procedure. Reprinted from CJS January 1969: (12) Pages(s) 83–88 by permission of the publisher. © 1969 Canadian Medical Association.)*

Atrial Switch Operations: Ake Senning and William Mustard

In 1957, Ake Senning (Fig 8, A and B), using flaps of autogenous atrial tissue performed a successful atrial switch in 1 patient (Fig 9) [10]. John W. Kirklin used the Senning operation at the Mayo Clinic and by 1961 had operated on 11 infants, with 4 survivors [24]. On May 16, 1963, Mustard (Fig 10) operated upon an 18-month-old girl who had previously undergone a Blalock-Hanlon operation. He first repaired the ventricular septal defect and then excised the entire atrial septum [25]. Then using an autologous pericardial baffle, Mustard performed an atrial switch operation (Fig 11). The girl made an uneventful recovery and developed normally. The authors of the present article had the pleasure of seeing and talking to this patient on May 27, 2001, during the opening ceremony of the Third World Congress of Pediatric Cardiology and Cardiac Surgery in Toronto. She continues to do well as of this writing and has two children.

A brief biographical note on Senning and Mustard seems appropriate.

Ake Senning (1915–2000) was born on September 14, 1915, in Rattwik, Sweden, to the family of a physician. In 1944, Senning received his medical degree after studying in Uppsala and Stockholm. In 1944, Dr Clarence Crafoord (1899–1984) carried out the first successful operation on a patient with coarctation, and 12 days later he did the second. The news of Crafoord's successful operations influenced Senning decision to become a cardiac surgeon. From 1948 to 1956, Senning trained under Crafoord at the Sabbatsberg Hospital in Stockholm. It is during this time that he and engineer Rune Elmquist developed a totally implantable pacemaker (Fig 8, B). From 1956 to 1961, Senning ran the Department of Experimental Surgery and was consultant surgeon at the Karolinska Hos-

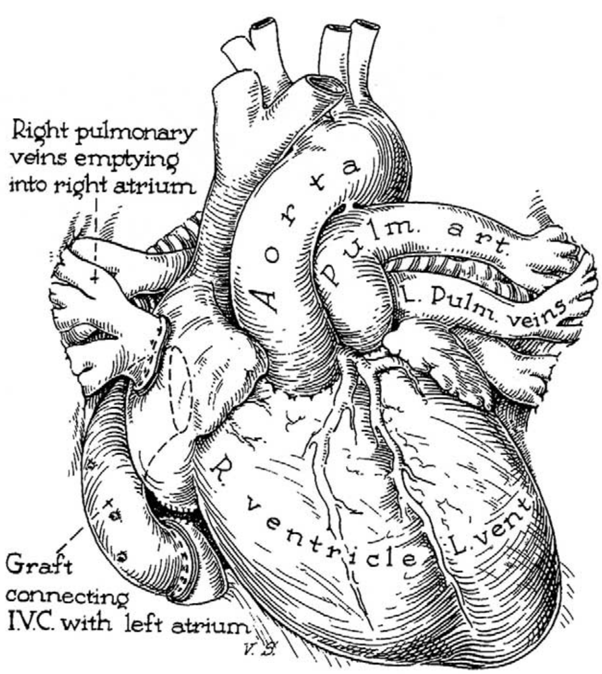

*Fig 5. Baffes operation. Diagram of completed transfer of the inferior vena cava (*IVC*) and right pulmonary veins. (*L. Pulm. veins *= left pulmonary veins;* L. vent. *= left ventricle;* Pulm. art. *= pulmonary artery;* R. ventricle *= right ventricle.) (Reprinted from Baffes TG, Surg Gynecol Obstet; 1956;102:227–33 [16], with permission.)*

MISCELLANEOUS

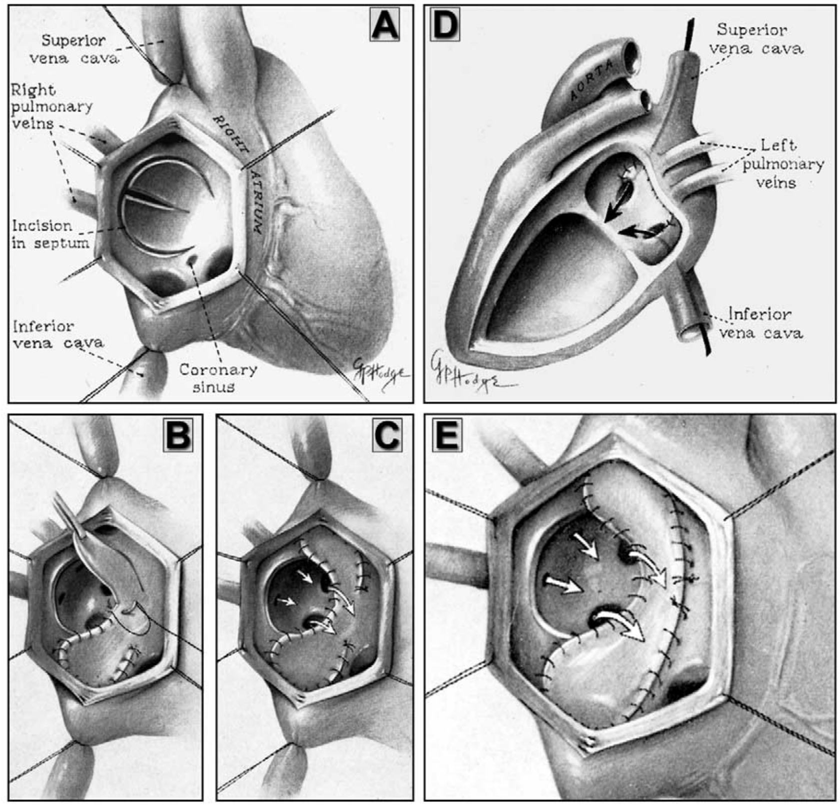

Fig 6. Albert operation. View from right atrium with surgically created bilobed flaps of interatrial septum (A). Each wing of the flap is rotated away from the central incision to cover the orifices of the vena cavae; the straight sides of the flaps, resulting from the central incision, are sutured to the posterior wall of the left atrium (B). View from right (C) and left (D) atrium after completion of the operation. If the septum is competely excised, a plastic graft could be used instead (E). The arrows *represent blood flow. (Reprinted from Albert HM, Surg Forum; 1954;5:74–7 [18], with permission.)*

pital. On October 8, 1958, he performed for the first time a total implantation of a cardiac pacemaker in a 43-year-old patient with complete heart block. From 1961 to 1985, Senning was full professor and chairman of the Department of Surgery at the University of Zurich. In 1969, he performed the first heart transplantation in Switzerland. Senning died on July 21, 2000, several months before his 85th birthday.

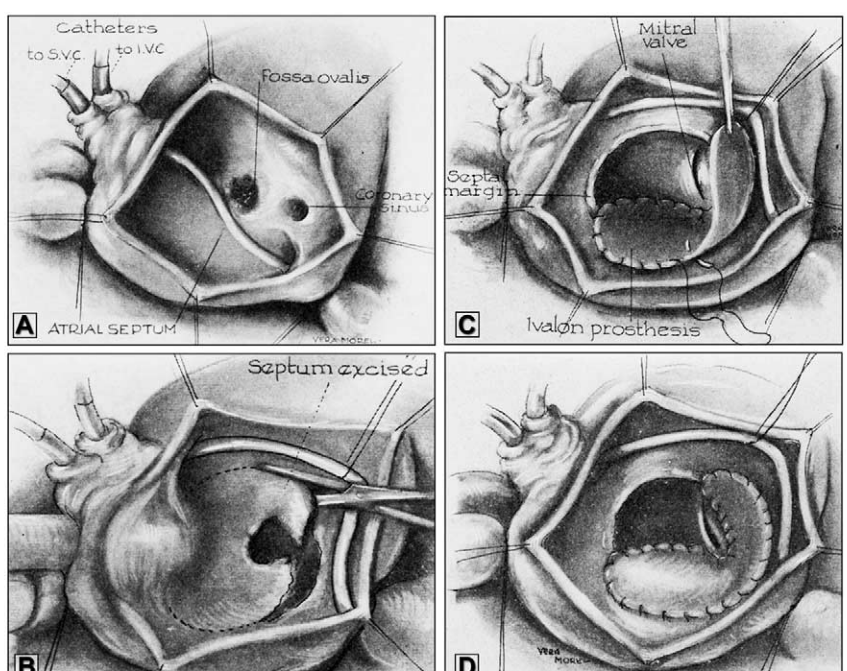

Fig 7. Creech operation. (A) View of the atrial septal defect in the region of the fossa ovalis and with caval catheters in place for extracorporeal circulation. (I.V.C. = *inferior vena cava;* S.V.C. = *superior vena cava.) (B) The atrial septum is completely excised thus creating a common atrial chamber; a prosthesis of polyvinyl sponge (Ivalon) is sutured over the orifices of the pulmonary veins (C) and over the tricuspid valve (D). (Reprinted from Creech O, et al, Surgery; 1958;43:349–54 [20], with permission.)*

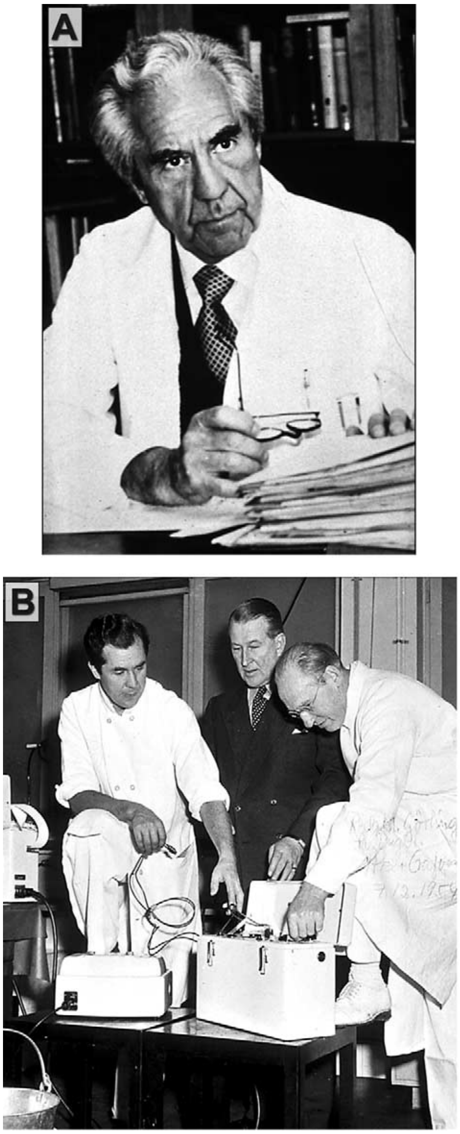

Fig 8. (A) *Ake Senning (1915–2000) around 1980s.* (B) *Ake Senning, Rune Elmquist, and Clarence Crafoord in 1954 (Courtesy of Dr Marko I. Turina).*

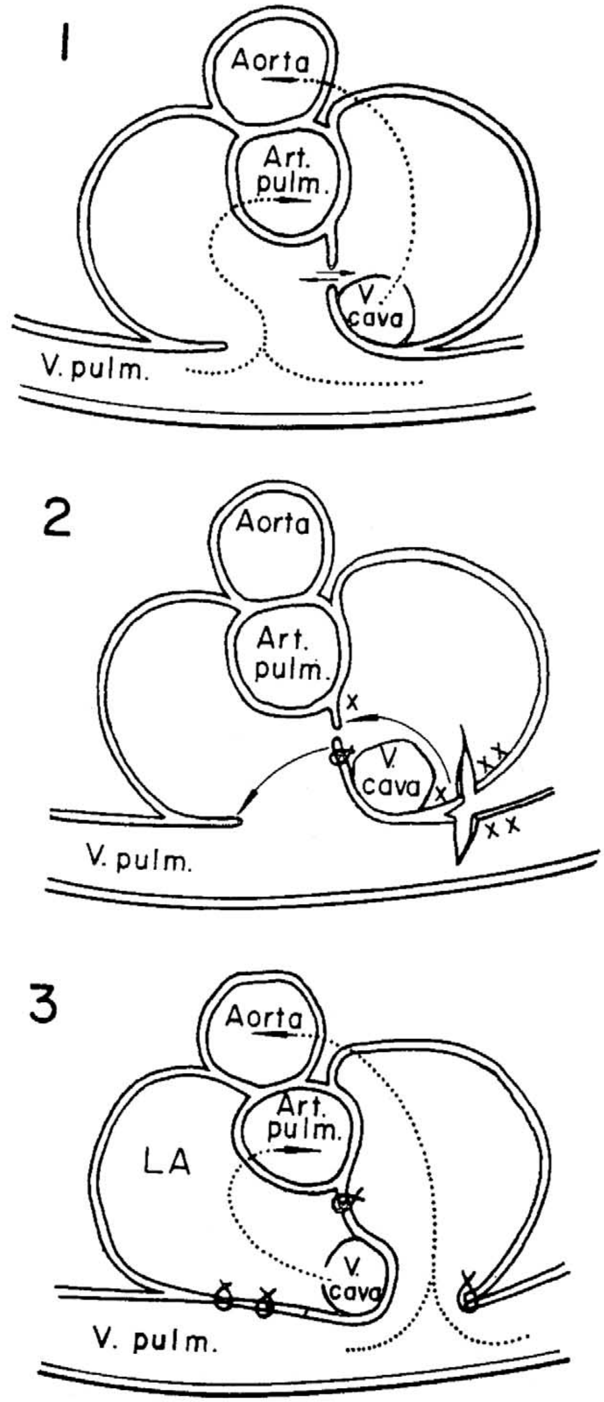

Fig 9. Senning operation. Diagrams of preoperative anatomy (1), surgical correction (2), and postoperative condition (3). The X *and* XX *indicate incisional edges to be sutures together. The* arrows *represent blood flow.* (Art. pulm. = *pulmonary artery;* LA = *left atrium;* V. cava = *vena cava;* V. pulm. = *pulmonary vein.) (Reprinted from Senning A, Surgery; 1959;45:966–80 [10], with permission.)*

William Thornton Mustard (1914–1987) was born on August 8, 1914, in Clinton, Ontario, Canada. His father was the principle in one of the best public schools in Toronto. He graduated from the University of Toronto Medical School in 1937 and joined the army in 1941. In 1944, at a casualty clearing station in Europe, he bridged a lacerated artery of a soldier's leg with a glass tube, managed the patient with heparin, and later replaced the glass tube with a vein graft. In 1947, Mustard spent a month observing the work of Blalock at the Johns Hopkins. This experience fueled his interest in cardiac surgery. From 1947 to 1985, Mustard served as orthopedic and cardiac surgeon at the Hospital for Sick Children in Toronto. In 1952 Mustard developed the first operation that was to bear his name—an iliopsoas tendon transfer—that allows children paralyzed by polio to be able to sit up. From 1957, Mustard devoted himself to cardiac surgery. Upon retirement in 1977, Mustard was awarded the Order of Canada in recognition of his achievements.

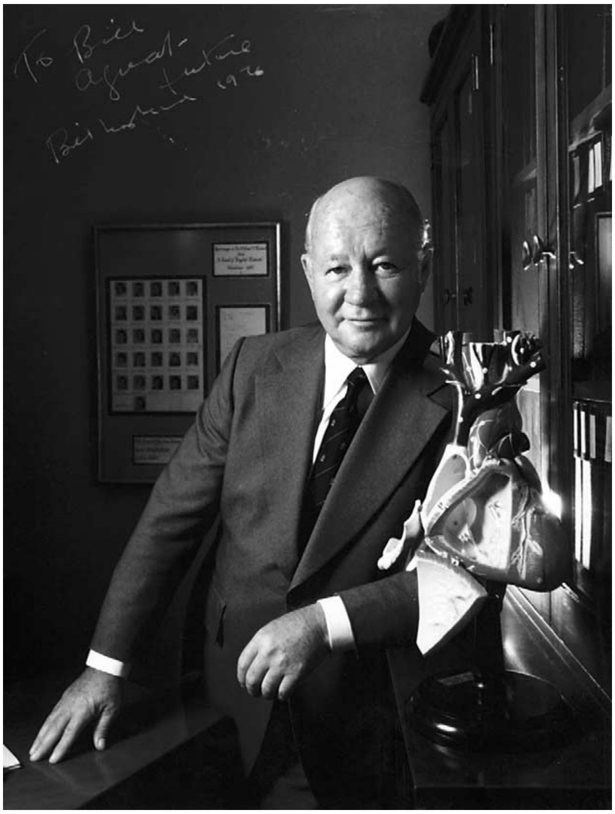

Fig 10. *William Thornton Mustard (1914–1987).*

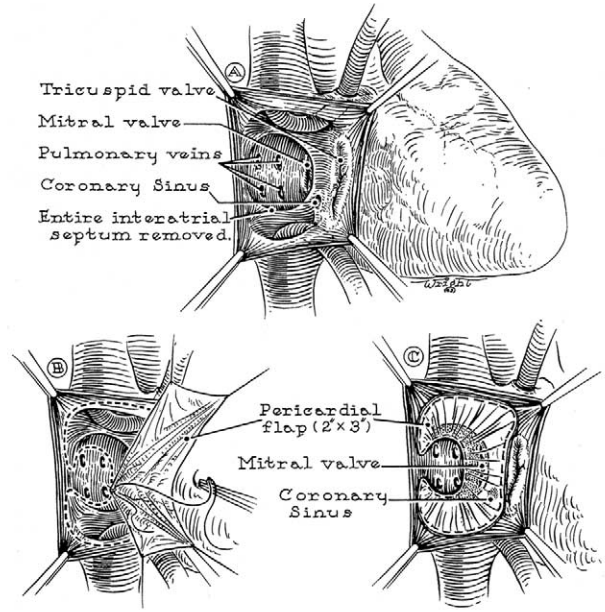

Fig 11. Mustard's operation. A pericardial baffle is used to reconstruct the atrial septum directing the venae cavae and coronary sinus flow to the mitral valve. The baffle allows pulmonary venous flow to tricuspid valve. (A) Intra-atrial anatomy. (B) Suturing of the pericardial flap. (C) Completion of the atrial switch. (Reprinted from Mustard WT, Surgery; 1964;55:469–72 [25], with permission.)

Ten years later, after having refused aortic valve surgery for himself a year earlier, Mustard suffered a massive heart attack while vacationing in Florida and died on December 11, 1987.

Senning Operation Versus Mustard Operation

Senning's operation was largely abandoned in 1964 after Mustard reported successful application of his less complex operation [26]. In 1966, Mustard presented a review of 25 patients who underwent his repair; all of them had normal arterial oxygen saturation and normal pulmonary and systemic venous pressures [27]. In 1966, Kirklin said in discussion after Mustard's presentation at the 44th annual meeting of the American Association for Thoracic Surgery in Montreal: ". . . I would agree that the Senning operation is obsolete and that the Mustard operation is clearly the treatment of choice for this condition. . .nearly half of the deaths, in our opinion, were on the basis of a low cardiac output. I think that the very small atria, which result from Senning procedure, may account for this high incidence of deaths from low cardiac output. Doctor Mustard's procedure results in much larger atria and may well significantly reduce the incidence of deaths from this problem" [21]. By 1966, Kirklin had accumulated perhaps the largest experience with Senning's operation. Impressed by Kirklin's work, Mustard was puzzled and said: "Dr. Kirklin is a tremendous technician. I don't know how on earth he has accomplished these operations. . ." [21]. With increased experience it soon became apparent that Mustard's operation was much easier to perform in small children. Mustard himself referred to Senning's operation as a "complicated procedure" that "although ingenious, is technically extremely difficult in the infant or small child" [27]. For the next decade, the Mustard operation was nearly universally employed (even by Senning) in younger children [28]. Subsequently, Mustard described an operative technique to provide a definitive repair after Baffes' procedure [29]. In his turn, Baffes applied the Mustard procedure for definitive correction of transposition after his operation [30].

It was not until 1975 that interest in the Senning operation was revived as a number of its potential advantages became more fully appreciated [31]. The Senning operation, with its avoidance of foreign material, seemed to provide a better growth and less venous obstruction in smaller children [28, 31]. For almost a decade many surgeons switched to the Senning operation.

In 1982, Jatene described the first successful arterial switch operation [32]. During the mid-1980s, the atrial switch operation was gradually replaced by Jatene's arterial switch. Subsequently, a survival advantage for patients with Mustard procedure and a rhythm advantage for patients with Senning procedure were demonstrated, but no important differences in functional class have been found [33, 34].

Present

From 1963 to 1998, 550 Mustard operations were performed at the Hospital for Sick Children in Toronto alone. After 1978 the arterial switch operation replaced the Mustard procedure for patients who had an associated ventricular septal defect, a group with poor results from atrial repair. Neonates with isolated transposition of the great arteries were changed to a protocol of arterial switch after October 1988. Survival of the first 547 patients after the Mustard operation at the Hospital for Sick Children is 64% at 25 years after surgery [35]. Both early and late survival after the Mustard operation for patients with isolated transposition of the great arteries (78% at 25 years) is better than survival for children with transposition of the great arteries and associated lesions, usually a ventricular septal defect [36, 37].

A long-term follow-up of neonates with complete transposition of the great arteries was recently published by the Congenital Heart Surgeons Society (CHSS) Data Center [38]. Between 1985 and 1989, the surgical management of neonates with complete transposition underwent a transition from atrial switch to arterial repair. The CHSS undertook a study to examine the intermediate outcomes and their associated risk factors in neonates repaired during the era of transition. Twenty-four institutions entered 829 neonates aged less than 15 days in a prospective study. Repair was by arterial switch (n = 516), atrial repair (n = 285) [Senning (n = 175), Mustard (n = 110)] or Rastelli (n = 28) operations. Survival estimates at 6 months, 5, 10, 15 years were 85%, 83%, 83%, and 81%, respectively [38]. Survival 15 years after transposition of the great arteries repair was good with most children functioning well, and results were best after an arterial switch operation. The recent study of quality of life in a cohort of 306 of these CHSS patients 11 to 15 years after transposition of the great arteries repair demonstrated that the quality of life and health status as perceived by these children themselves were excellent when compared with published data for normal children and were better after the arterial switch than after either atrial switch operation [39]. Currently, most patients with an atrial switch are now adults, lead active lives, but need ongoing expert care in an adult congenial cardiac center.

Future

In 1987, Senning wrote: "I think that the arterial switch—a real anatomical correction—will be the 'golden standard' in the near future and that the atrial switch will be used only for the few patients who are not suitable candidates for the arterial switch" [28]. This is what has happened!

As time passed, atrial repair of transposition of the great arteries has been almost entirely replaced by the arterial switch operation. However, the atrial switch operation is not completely obsolete. Neither is it a procedure of simply historical interest. Congenital cardiac surgeons need to be familiar with the procedure as there will be four situations where the atrial switch will be indicated in future:

The first indication is for infants with isolated transposition of the great arteries who first present after the neonatal period, the atrial switch operation is an alternative to two-stage arterial switch operation (namely, a preliminary pulmonary artery banding to prepare the left ventricle, followed after an interval by the arterial switch operation). The low risk of the atrial switch and favorable long-term results in infants with isolated transposition of the great arteries support its consideration in this situation. In contrast, infants with transposition of the great arteries and an associated ventricular septal defect that present after the neonatal period would not have lost their left ventricular hypertrophy and are managed with primary arterial repair, not an atrial switch.

The second indication for the atrial switch operation is palliation of patients with pulmonary vascular disease from an associated ventricular septal defect. The atrial switch provides preferential streaming of the blood flow and improves the arterial oxygen saturation in patients who would not tolerate ventricular septal defect closure and arterial switch [40].

The third indication for the atrial switch is for patients with congenitally corrected transposition in whom repair includes both a venous and arterial switch to create ventriculoarterial concordance or, in patients with associated pulmonary valve stenosis or atresia, an atrial switch combined with a Ratelli operation (namely, Ilbawi's operation, also known as "double switch" or "anatomical correction") [41, 42].

The fourth indication for the atrial switch procedure is for the rare anomaly of isolated ventricular inversion. In this situation there is atrioventricular discordance with ventriculoarterial concordance [43]. The atrial switch operation corrects the circulations physiologically and leaves the morphologic left ventricle supporting the systemic circulation. An arterial switch operation is contraindicated because of the presence of ventriculoarterial concordance. This is a very rare anomaly. Of 550 patients, who underwent the Mustard operation in Toronto, only 1 had isolated ventricular inversion.

The atrial switch operation resulted from the cumulative effort of many surgeons. Their creativity is a source of inspiration for all of us. Their success was a matter of life and death to their patients. Their work made a difference. It made our world a bit better and initiated enormous improvements in the care of the patients with congenital heart diesease. We conclude with the words of Dr Baffes, who wrote in 1987: "History tends to judge a human being on two counts: the use made of the time allotted to him and what, if anything, he leaves behind for the generations that follow" [22]. The surgeons who developed the atrial operations for transposition left behind them the inspiration for the subsequent impressive advancements in congenital heart surgery.

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