Mustard Operation

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The natural history of infants with complete transposition (TGA) is rapid deterioration and death within the first few months of life. The few babies who do survive beyond the first year of life usually have an associated ventricular septal defect (VSD) and subsequently deteriorate with progressive pulmonary vascular disease.

Intervention to improve the outlook for newborns with TGA began with surgical resection of the atrial septum.[1] The enlarged atrial defect improves mixing of the pulmonary and systemic circulations, thereby increasing the systemic oxygen saturation. Balloon atrial septostomy, as described by Rashkind and Miller in 1966,[2] replaced surgical septectomy. The balloon catheter technique continues to provide effective emergency palliation for the newborn with TGA.

In the early 1950s, a number of surgeons, including Mustard,[3] attempted arterial repair of TGA in infants, all without success until Jatene's report in 1978.[4] In 1954, Albert[5] proposed a surgical technique to transpose the venous inflow within the atria to correct TGA. This concept of atrial repair of TGA was first successfully used by Senning in 1958.[6] He devised an ingenious technique of creating atrial flaps to redirect (transpose) the pulmonary and systemic venous inflow. Senning's technique was difficult for other surgeons to duplicate, however.

Mustard, frustrated by his failure with the arterial repair, devised a pericardial baffle technique of atrial repair.[7] His concept evolved from his experience with reoperating on children who had arterial desaturation after repair of an atrial septal defect (ASD). In that early era of open heart surgery, children with ASDs were repaired without cardiopulmonary bypass. During a 3-minute occlusion of both vena cavae, the ASD was suture closed, occasionally inadvertently leaving a leak at the caudal end that diverted inferior vena cava (IVC) flow into the left atrium and produced systemic desaturation. Mustard reasoned that both vena cavae could be deliberately diverted to the left atrium while allowing the pulmonary return to enter the right atrium, thereby correcting the circulation in patients with TGA. The Mustard baffle operation was a simple, reproducible, and elegant technique of transposing venous return. It was easier to learn than the Senning operation, and became the standard repair in most of the world.

Indications for the Mustard Operation in 1998

The atrial repair of TGA has been almost entirely replaced by the arterial switch operation. However, there are three situations were the Mustard operation may be indicated:

1. For infants with isolated TGA who first present after the neonatal period, the Mustard operation is an alternative to the two-stage arterial switch operation, ie, a preliminary pulmonary artery banding to prepare the left ventricle, followed after an interval by the arterial switch operation. The low risk of a Mustard operation and favorable long-term results in infants with isolated TGA support its consideration in this situation.

In contrast, infants with TGA and an associated VSD that present after the neonatal period would not have lost their left ventricular hypertrophy and are managed with primary arterial repair, not a Mustard operation.

2. The second indication for the Mustard operation is for palliation of patients with pulmonary vascular disease from an associated VSD.

3. The third indication for a Mustard operation in the present era is for patients with congenitally corrected transposition in whom repair includes both a venous and arterial repair to create ventriculo-arterial concordance, or the so-called "double switch" operation.[8]

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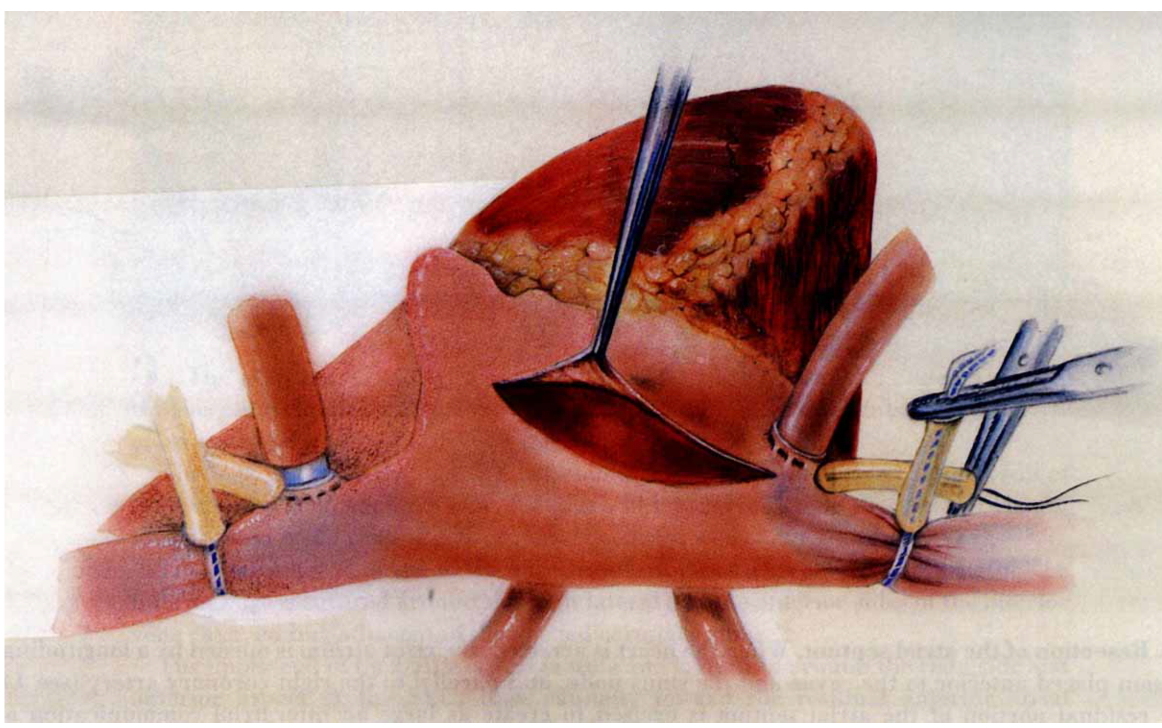
SURGICAL TECHNIQUE

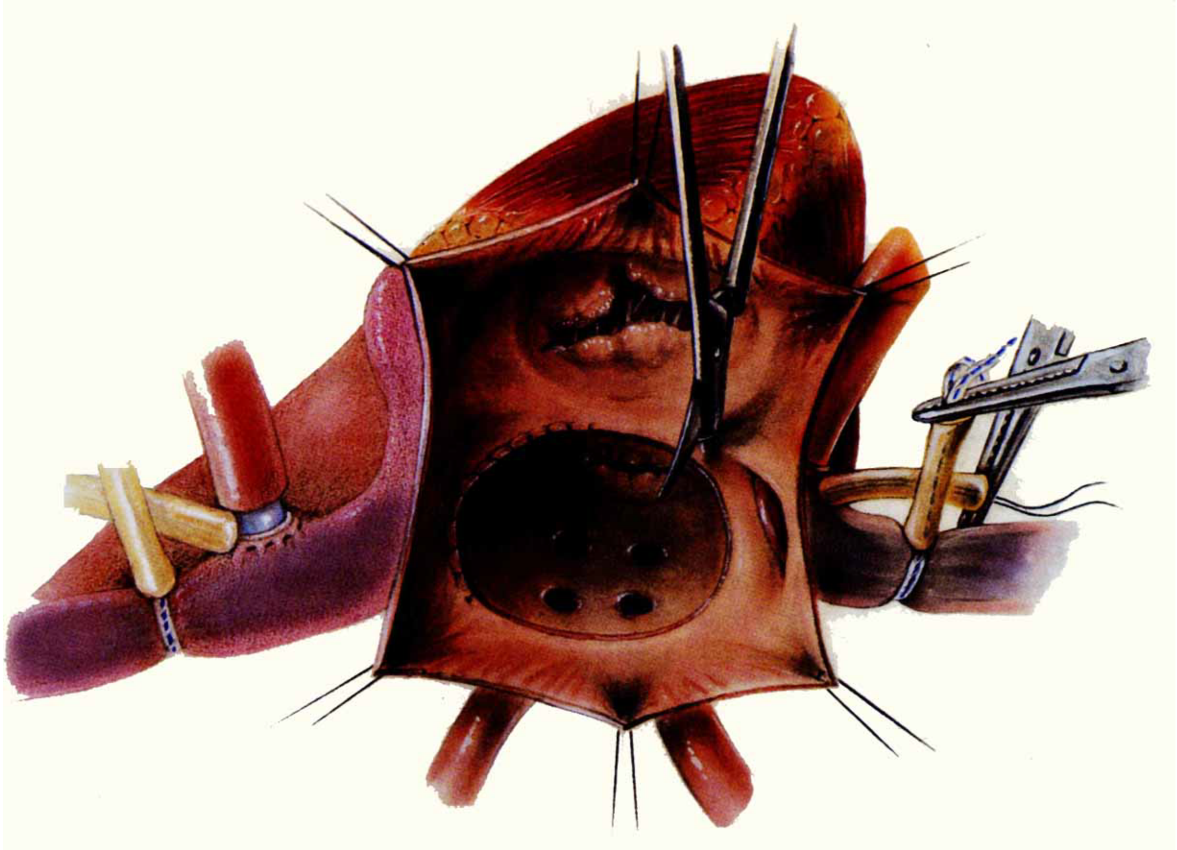
Preparation of the patient. Preoperative assessment should clarify the associated cardiac lesions and establish that the patient is free of concurrent illness.

The Mustard operation is performed under general anesthetia with central venous, arterial, electrocardiographic, and urinary output monitoring. A left-atrial (morphological right atrial) pressure catheter is placed after repair for routine postoperative monitoring. Intraoperative transoesophageal echocardiography should be performed as the patient is weaned from bypass, to confirm that the pericardial atrial baffle is not causing stenosis of either of the venous pathways and is free of a suture line leak. Temporary bipolar pacing wires are placed on the right atrium and ventricle for postoperative monitoring care.



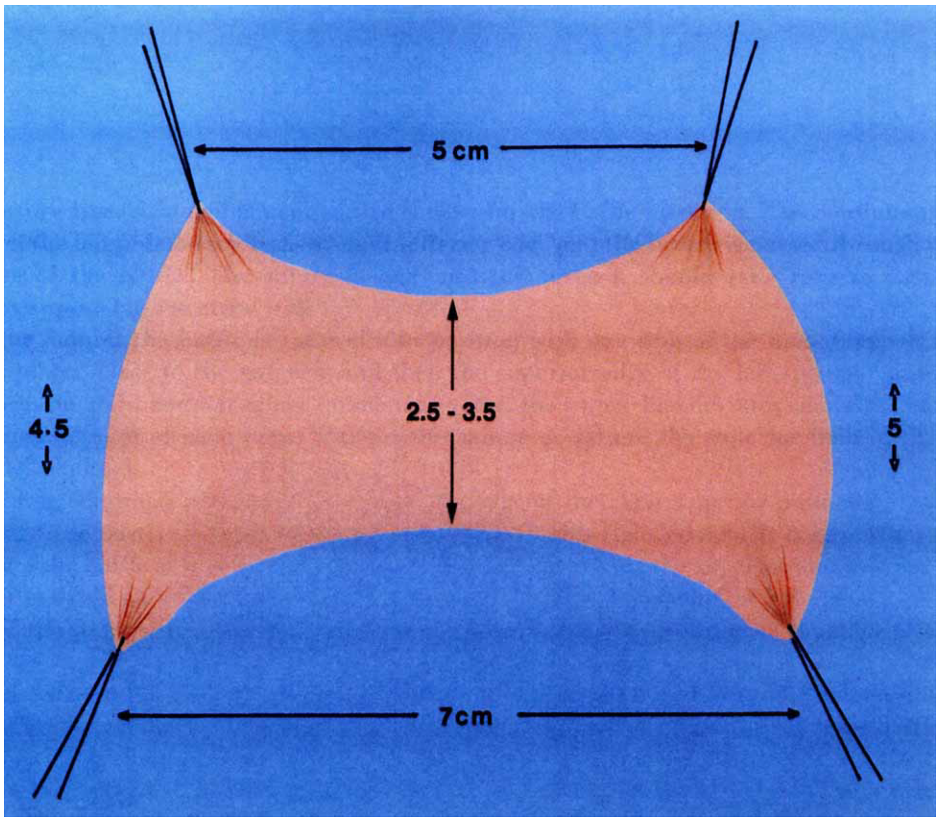
1 Although most of the children in our series underwent repair during a short period of hypothermic circulation arrest, we now use standard cardiopulmonary bypass with bicaval cannulation. The superior vena cava (SVC) is cannulated directly through its anterior wall, 1 to 2 cm cephaled to the atrium (see figure). The IVC is cannulated through a purse-string incision adjacent to the IVC, or on the anterior wall of the IVC.

The heart is arrested with warm-induction (35°C to 37°C) blood cardioplegia, and once arrested, is cooled by the cardioplegia to 15°C. Topical cooling of the heart with a water-circulation cooling jacket will maintain a low myocardial temperature and repeat doses of cardioplegia are usually unnecessary. Terminal warm-blood cardioplegia allows controlled recovery of the heart and facilitates the important maneuver of air removal from the anteriorly located systemic, morphological right, ventricle.



2 Resection of the atrial septum. When the heart is arrested, the right atrium is opened by a longitudinal incision placed anterior to the cavae and the sinus node, and parallel to the right coronary artery (see 1). The residual portion of the atrial septum is excised to create as large an interatrial communication as possible (see figure). Resection of atrial tissue adjacent to the sinus (SA) and atrio-ventricular (AV) nodes is limited to avoid direct trauma to these structures. The artery to the SA node is usually located at the base of the anterior wall of the septum medial to the SVC-right atrium junction, and resection of the atrial wall in this area should be limited. The atrial septum adjacent to the right pulmonary veins can be excised deeply. The superior wall of the coronary sinus is incised into the left atrium as far as the posterior leaflet of the mitral valve to create a larger interatrial communication.

The exposed atrial muscle is re-endothelialized by a suture line, which re-approximates the endothelial edges of the resection. We use an absorbable suture to facilitate growth.



3 **The pericardial baffle.** The size and shape of the pericardial baffle is critical to the success of the Mustard operation. The baffle is initially rectangular and the 5 × 7 cm size was designed for a child aged 9 months to 2 years.

The 7-cm length is the distance between retraction sutures placed at each end of the (initially) rectangular baffle. Its edge will be sutured along the anterior side of the left pulmonary veins and over to the posterior aspect of each vena cavae.

The opposite side of the baffle and the inferior end are each 5 cm in length. The inferior edge is sutured around the right lateral and the anterior sides of the inferior vena cava, ending adjacent to the opened coronary sinus.

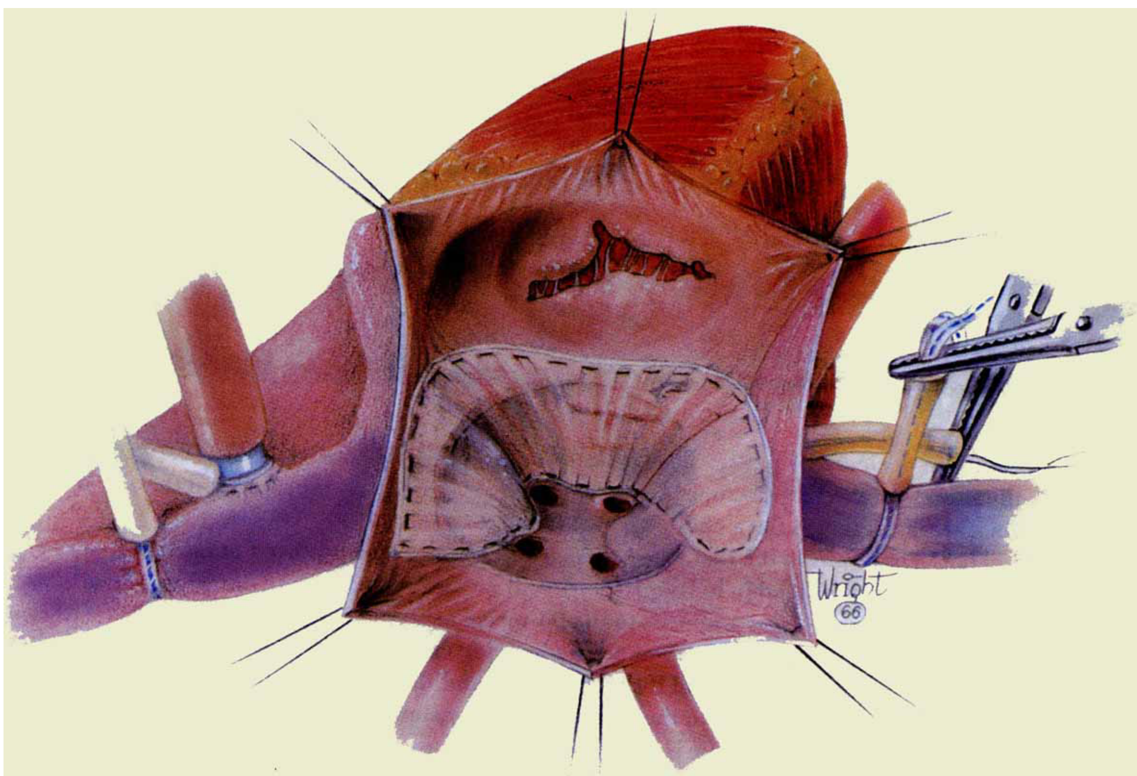
The upper end of the baffle is 4.5 cm wide and is sutured around the right side and anterior aspect of the SVC, then caudally toward the residual anterior atrial septum. The fourth side of the baffle, 5 cm in length, is sutured to the residual anterior base of the atrial septum.

The rectangular shape of the baffle is modified before insertion as follows: the central portion of the baffle is cut to a narrow waist measuring 3 cm. Both the 7 cm and the anterior 5 cm edge of the baffle are trimmed in a smooth convex curve to create the narrow central portion after the previously stated dimensions are established.

In addition, each end of the baffle is rounded slightly, creating a convex curve to facilitate rounding of the baffle around each cava.[9] The rounding of the end adds about 0.5 cm to the baffle at each end.

Size modifications. For younger or older patients, the proportions of the pericardial baffle remain the same, but the size varies, although not very much.

For a neonate, the basic 5 × 7 cm size would decrease to 4 × 6 cm, with the central portion narrowed to 2.5 cm. The SVC end of the baffle would be 3.5 cm, and each end is rounded as described previously. For an adult, the baffle size increases to 6 × 8 cm, with a central waist of 3.5 cm.



4 See legend on opposite page.

4 Baffle suture line. A monofilament suture is used for the baffle insertion. The continuous suture line begins between the left pulmonary veins at their anterior edge and the suture is passed through the center of the 7-cm edge of the baffle. The suture is tied and the surgeon should take care to ensure that the pericardium is apposed to the atrial wall.

The superior limb of the baffle is sutured into position with one arm of the monofilament suture. The suture line is placed close to the anterior and then the superior edge of the left superior pulmonary vein orifice. Between the right and left upper pulmonary veins, the suture line deviates caudally to an imaginary line joining the midpoint of each vein. Without this minor deviation, the superior limb of the baffle may narrow the SVC channel.

The suture line continues cephalad to the superior edge of the right superior pulmonary vein and then cephalad toward the posterior point of the junction of SVC with right atrium. It is important to place the superior limb of the baffle as high (ie, cephalad) as possible to separate the superior and inferior limbs of the baffle in order to avoid a narrow channel for the pulmonary veins, especially the left veins.

The first of the four corners of the baffle should lie at the posterior-inferior junction of the SVC-RA junction.

The superior edge of the baffle, with its slightly convex edge, is sutured around the lateral and anterior edge of the SVC as far cephalad as possible. The sutures should be placed wider apart in the baffle than in the atrial tissue to gather the pericardium, thereby promoting bulging of the baffle around the SVC orifice. There are atrial trabeculae in this area and the SA node is located at the base of the superior extension of the crista terminalis.[9]

Placing the suture line high, ie, into the cavo-atrial junction above the trabeculae can avoid both the risk of damage to the SA node and a baffle leak around a trabecula adjacent to the suture line.

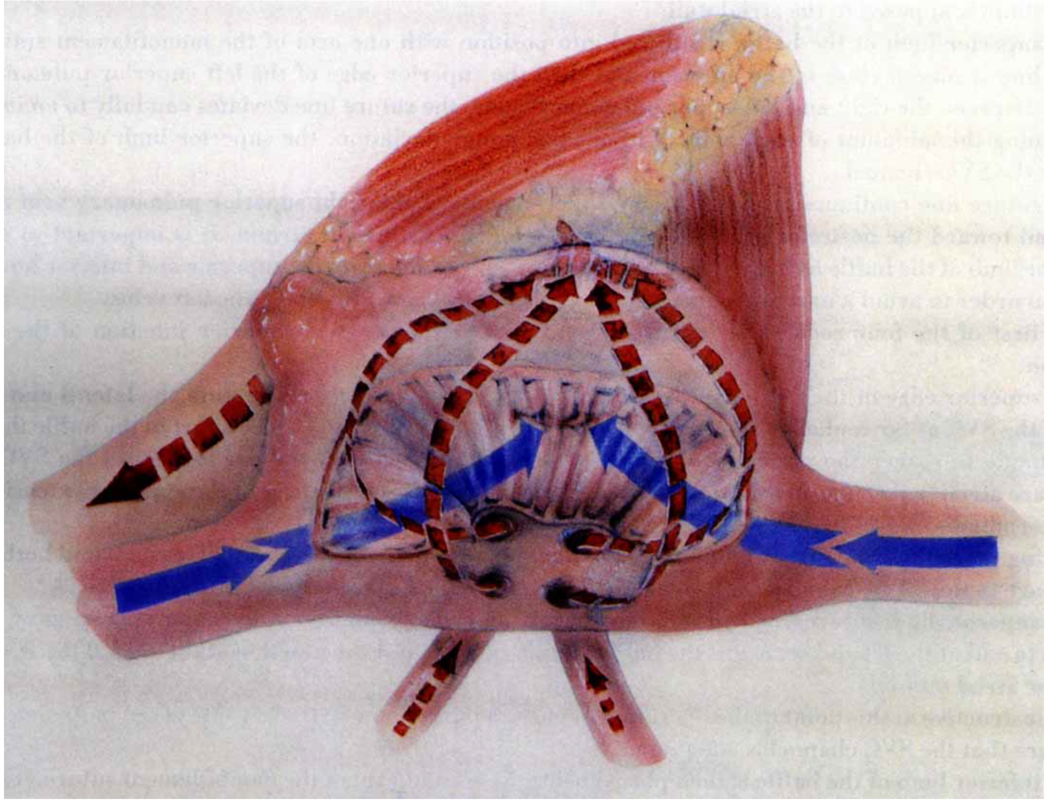
The superior-limb suture line continues anteriorly and then caudally on the superior aspect of the right atrium to end at the second corner of the baffle, which should lie at the junction of the roof of the RA with the anterior atrial septum.

It is instructive at this point in the operation to pass an instrument on the left side of the baffle into the SVC to ensure that the SVC channel is adequate.

The inferior limb of the baffle is then placed using the second arm of the monofilament suture. The suture line is initially along the anterior edge of the left inferior pulmonary vein. At the inferior edge of the left inferior pulmonary vein, suturing follows an imaginary line drawn from that point to the lateral posterior junction of the IVC and RA. This portion of the suture line must allow for as wide an angle as possible between the inferior and superior baffle limbs to ensure an adequate pulmonary venous channel. The third corner of the baffle should lie 1 cm proximal to the lateral-posterior junction of the IVC and RA.

The inferior side of the baffle is sutured to the lateral and then anterior edge of the IVC. As for the SVC, the pericardium is gathered onto the IVC by taking wider suture bites in pericardium than in caval tissue. The suture continues medially from the anterior-medial edge of the IVC toward the splayed open orifice of the coronary sinus where the fourth corner of the baffle should lie. In the opened orifice of the coronary sinus, the suture line is placed 1 to 2 mm into the coronary sinus and very superficially to avoid injury to the AV node. It is instructive to probe the IVC channel to ensure an adequate pathway.

The fourth side of the baffle is then sutured to the base of the anterior margin of the atrial septum, reaching the superior limb suture at the roof of the right atrium. This final portion of the suture line is the only section that follows the plane of the natural atrial septum.



5 When the baffle is complete, the caval tapes should be released to fill the morphological left atrium and ensure that there are no suture line leaks. In addition, distention of the baffle allows the surgeon to assess the adequacy of the pulmonary venous channel (see figure). If the limbs of the baffle protrude to the point of encroaching on the pulmonary venous drainage, interrupted plication sutures to one or both limbs can be inserted, taking care to avoid excessive narrowing of the systemic channels.

The right atriotomy is then closed directly, and great care is required to ensure complete evacuation of all air from the pulmonary veins and the anteriorly positioned systemic ventricle.

Results

From 1963 to 1998, we performed 550 Mustard operations. The arterial switch replaced the Mustard operation for patients with an associated VSD after 1978 and for neonates with isolated TGA, after October 1988.

Survival of the first 547 patients is shown in Figure I. Both early and late survival after a Mustard operation is better for patients with isolated TGA[10-11] compared with survival for children with TGA and associated lesions, usually a VSD[12] (Fig II). The association of poor outcome with associated VSD persists into adulthood.

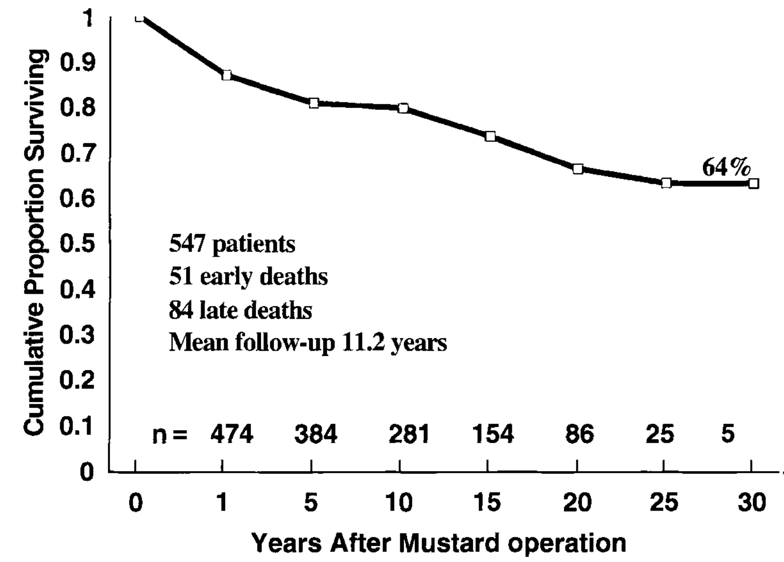


Fig I. Actuarial survival (Kaplan-Meier) in 547 patients after a Mustard operation at the University of Toronto. Survival 25 years after operation is 64% n = 25, the number of patients entering each time period indicated on the horizontal axis.

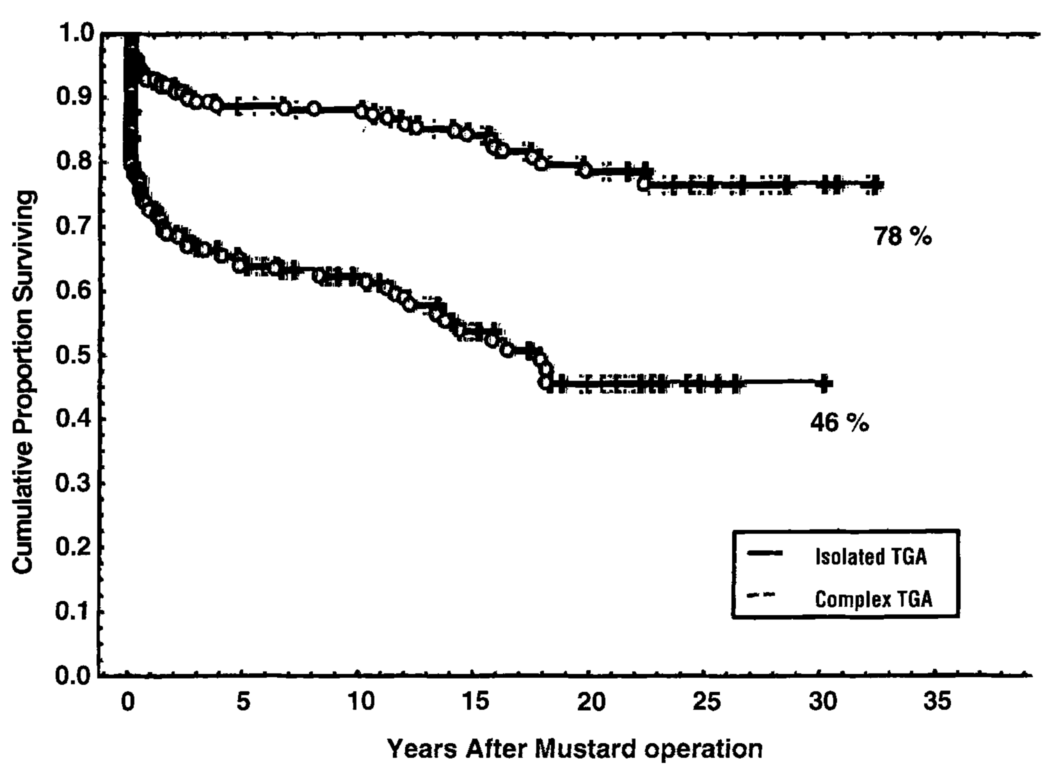


Fig II. Survival of patients with isolated transposition after a Mustard operation (n = 375) is 78% at 25 years. For patients with complex transposition (n = 172), early and late survival is significantly lower (P .001).

Among our patients that reach the age of 18 years with isolated TGA, chance survival to age 41 years is 91% by actuarial analysis. In contrast, those with a VSD have only a 38% chance of survival to age 30 years.

An analysis of our late results by Gelatt et al[13] clarifies the issues regarding ventricular dysfunction and arrhythmia. Severe RV dysfunction (grade 4) was evident in 7% of our patients and is associated with poor survival. Lesser degrees of ventricular dysfunction were not discriminating in predicting long-term survival. Independent risk factors for late death are: earlier date of operation, the presence of a VSD, and perioperative arrhythmia.

During follow-up, there is a progressive loss of sinus rhythm and an increasing prevalence of atrial, and to a lesser extent, ventricular arrhythmias. Only 40% of our patients are in sinus rhythm 20 years after a Mustard operation and 20% have developed atrial flutter. Multivariable analysis showed that sudden death, which accounts for 40% of late deaths, is associated with smaller patient size at the time of operation, the presence of perioperative arrhythmia, and the presence of AV block.

Conclusion

The Mustard operation dramatically changed the outlook for babies born with TGA. Although it has been replaced by the arterial switch operation, the Mustard operation is a remarkable contribution to the development of cardiac surgery and to the care of children with congenital heart disease. Most survivors of the Mustard operation are young adults. They require ongoing follow-up and are at risk for loss of sinus rhythm, the onset of atrial arrhythmia, and sudden death.

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