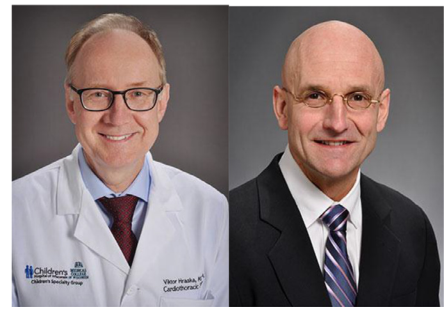
Anatomic Repair of Corrected Transposition of the Great Arteries: The Double Switch

Viktor Hraska, MD, PhD, and Ronald K. Woods, MD, PhD

The long-term outcome of patients with corrected transposition of the great arteries and associated lesions after physiologic repair is uncertain. Anatomic correction, utilizing the morphologic left ventricle as a systemic pumping chamber and the mitral valve as the systemic atrioventricular valve, is considered the preferred method, especially for patients with either tricuspid valve regurgitation, with Ebstein's malformation of the tricuspid valve, or with right ventricular dysfunction. The double switch employs both an atrial switch and arterial switch to "correct" the atrioventricular and ventriculoarterial discordance. Associated lesions are also repaired. The best outcomes with double switch are achieved with patients in the first few years of life even if reconditioning of morphologic left ventricle is required. However, the long-term function of the conduction system, the aortic valve, and the ventricles is variable and requires close surveillance.

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 22:57–60

Keywords: Corrected transposition of the great arteries, Physiological repair, Anatomical repair, Double switch procedure



Viktor Hraska, MD, PhD; Ronal K. Woods, MD, PhD.

Central Message

The best outcomes with double switch are achieved with patients in the first few years of life even if reconditioning of the morphologic left ventricle is required.

Treatment of congenitally corrected transposition of the great arteries (ccTGA) is multifaceted and includes observation with close follow-up, medical therapy for heart failure, pacemaker insertion, and surgery [1,2]. Despite isolated case reports of preserved morphologic right ventricular (mRV) function in advanced age, there is general agreement that the fundamental problem is the inability of the mRV and tricuspid valve (TV) to support the systemic circulation long term [2]. To address this fundamental issue, techniques have been developed to both correct the associated defects and restore a concordant circulation with the morphologic left ventricle (mLV) and mitral valve (MV) supporting the systemic circulation [3]. Double switch (DS) represents one type of anatomical correction in which the atrioventricular discordance is corrected by an atrial switch (Senning or Mustard), and the ventriculoarterial discordance is corrected by an arterial switch operation. With technical aspects of the procedure well established, DS is safe and provides encouraging mid-term survival and functional benefits, even in patients requiring preoperative training of the mLV [4]. However, certain complexities and potential complications may temper the enthusiasm for pursuing this strategy of care.

Division of Congenital Heart Surgery, Department of Surgery, Herma Heart Institute, Medical College of Wisconsin, Milwaukee, Wisconsin

Conflicts of Interest and Funding: The authors have nothing to disclose with regards to commercial support.

Address correspondence to: Viktor Hraska, MD, PhD, Division of Congenital Heart Surgery, Department of Surgery, Herma Heart Institute, Medical College of Wisconsin, 9000 W. Wisconsin Avenue, B730, Milwaukee, WI 53226. E-mail: vhraska@chw.org

DECISION-MAKING AND MANAGEMENT STRATEGY

Decision-Making

The indications and the timing of the operation depend on the clinical state and age of the patient. Provided both ventricles are balanced and properly developed, the DS procedure is indicated if the mLV is properly trained for systemic function and there is no obstruction of the mLVOT (LVOTO), or obstruction is surgically resectable. Resectable LVOTO includes membranous stenosis, aneurysmal dilation of the fibrous tissue derived from the interventricular component of the membranous septum, or accessory tissue from atrioventricular (AV) valves, or a stenotic, but well-formed pulmonary valve (Video 1).

Primary DS with unrestrictive ventricular septal defect (VSD) is usually performed between 3 and 6 months of age. If pulmonary artery banding (PAB) is necessary to control congestive heart failure due to a large VSD, the DS is performed between 6 and 18 months of age. If there is no VSD and a PAB is placed

1092-9126/$–see front matter
https://doi.org/10.1053/j.pcsu.2019.02.003

to control heart failure due to TV regurgitation, the DS is considered when the mLV is properly trained for systemic function, which usually takes more than a year.

Surgical Management

Training of mLV by PAB

As a general rule, PAB is required in patients with intact ventricular septum beyond the third month of age to prepare the mLV. General principles of mLV training are elaborated elsewhere.

Currently, we prefer the concept of enhanced mLV training [5], which includes a balance of both increased afterload and preload, detailed below:

1. placing a "loose" PAB achieving 50–60% of systemic pressure in the mLV;
2. unloading the mRV and increasing the mLV preload by creating an atrial septal defect (ASD—usually at least 10 mm in diameter).

This approach seems to simplify postoperative management and provides gradual interval training of the mLV. The procedure is done on cardiopulmonary bypass with a short aortic cross-clamp time. Direct mLV pressure measurements and transesophageal echo guide PAB adjustment based on the position of the interventricular septum and ventricular and tricuspid valve function [4].

VSD Closure

Closure of the VSD requires placement of sutures on the mRV side of the septum, particularly in the region of the conduction system. Our practice is to place all sutures on the mRV side. Approach to the VSD can be through the MV, mR-ventriculotomy, or aortic valve. The latter may prevent undue tension on the crux of the heart, but should carefully preserve the function of the aortic valve. In the event that heart block occurs, one should consider biventricular pacing, as there are several reports of progressive mLV dysfunction in those with mRV pacing alone [6].

Modified Senning Operation

The technique is adapted to the specific atrial morphology of the ccTGA with underdevelopment of the free right atrial wall, especially in situs solitus with mesocardia or dextrocardia, or in situs inversus with levocardia [4]. Modification of the original technique includes:

1. preservation of the superior vena cava-right atrium (SVC-RA) pericardial reflection to avoid damage of the sinus node;
2. cryoablation of cavo-mitral isthmus [7];
3. complete resection of the interatrial septum with effective cut off of the superior aspect of the limbus of the fossa ovalis to avoid baffle obstruction from the SVC to the TV. At the same time, the trapezoid-shaped patch, which is used for the posterior wall of the systemic baffle, should be seated low beneath the SVC-RA junction;
4. unroofing and incorporating the coronary sinus to the systemic venous pathway;
5. use of autologous pericardium in situ to create the pulmonary venous atrium.

The modified Senning operation is simple and highly reproducible regardless of situs or position of the ventricular apex. It enables repair at an early age, provides adequate capacity of the pulmonary venous atrium, preserves optimal geometry of the mitral valve, minimizes risk to the sinus node, and preserves the coronary sinus for electrophysiologic studies or intervention (Video 1).

Arterial Switch Operation

While the goal and technique of arterial switch operation for ccTGA is the same as for TGA, several technical points should be noted [8,9]:

1. standard coronary transfer principles apply, albeit with mirror image position of the coronaries;
2. thorough mobilization of the anterior button (LAD, Cx) is necessary as this button requires a bit more distance to rotate to the neo-aorta compared to the more posterior button;
3. the patch material of the reconstructed neo-pulmonary artery is left long to avoid anastomotic tension—translocation of the pulmonary artery bifurcation leftward may help reduce tension and/or avoid coronary compression;
4. leaving a relatively long neo-pulmonary artery (by transecting the aorta well above the sinotubular junction) again allows for more flexibility in reconstruction;
5. the LeCompte maneuver is rarely used (only for anteropostero great arterial relationship);
6. coronary transfer should not alter the diameter of the sinotubular junction, and annuloplasty (if age appropriate) may help prevent the development of aortic insufficiency (Video 1) [10].

Tricuspid Valve Surgery

Restoring the mRV and TV to the low pressure pulmonary circulation invariably results in improvement of both ventricular and valve functions, even in the context of valve dysplasia. Therefore, concomitant TV repair is rarely needed at the time of anatomic correction [4,9].

LONGER TERM OUTCOMES AND COMPLICATIONS

The long-term outcome after physiologic repair is unsatisfactory with a 20-year survival of 50–80% depending on the particular anatomy and physiology at the time of surgery [11–13]. Cause of death includes sudden death, myocardial failure, reoperations, arrhythmias, and infection. The status of the systemic mRV is a very strong determinant of long-term outcome [14,15]. Mid-term reoperation-free survival is between 50% and 80% with the worst outcomes seen in patients with TV replacement, particularly if there is significant mRV dysfunction at the time of valve replacement [12,13,15]. Survivors of physiologic repair demonstrate markedly reduced exercise capacity [16].

The only exception is a patient with native well-balanced pulmonary stenosis, which has a protective effect on TV and mRV function [17]. In patients not suitable for anatomic correction, prophylactic PAB with limited atrial septal communication might be useful as a palliation to mimic the favorable history of patients with ccTGA and limited LVOTO.

The survival and functional benefits of anatomic repair have been reported by several authors, particularly in patients with TV regurgitation [4,13]. Ten- to 15-year survival for the double switch has been reported as 78–93% [4,8,9,13]. Event-free survival ranges between 70% and 91% at 10 years [4,13,18]. Reintervention on the Senning pathways ranges between 5% and 12% [4,8,9,19]. Need for some form of reoperation is possibly inherent to any type of repair for ccTGA. Lim et al, in a comparison of 123 patients with physiologic repair to 44 patients with anatomic repair, reported 15-year freedom of reoperation of 35% for physiologic repair and 46% for anatomic repair (statistically insignificant) [13].

The main concern after DS procedure is deterioration of mLV function which has been reported to occur in 15–20% of patients at 20 years of follow-up. The risk is increased in patients with late neo-aortic and/or mitral insufficiency, heart block, and preoperative heart failure [6,8,18,19]. However, one single-center report demonstrated 93% freedom from significant ventricular dysfunction at 20 years of follow-up for anatomic repair vs 19% for physiologic repair [13]. Hraska et al reported 93% freedom from death, heart transplant, and reduced mLV function at 15 years of follow-up in a series of 63 consecutive patients (37 patients had DS) who underwent anatomic repair [4]. These outcomes are superior to physiologic repair.

The risk of ventricular dysfunction appears to be highest in patients requiring mLV training. Age at initiation of training is a critical variable as training after 2 years of age has been demonstrated as a risk for mLV dysfunction, whereas patients trained earlier remained free of dysfunction [20]. Zartner et al reported encouraging experience with enhanced mLV training [5]. In fact, this group reported no difference in mLV function after DS in patients with and without the need for training [4]. Enhanced mLV training may decrease the strain on mLV, thus potentially improving long-term function. In patients with a significant shunt at the atrial level (Qp/Qs > 1.5 and a defect size of at least 10 mm), it is known that the additional volume load will create a functional gradient across the pulmonary valve as the narrowest point in the pulmonary circulation [21]. As long as the diastolic function of the mLV is preserved, the atrial shunt increases the transpulmonary gradient during physical activity, which, in effect, creates a form of intermittently elevated mLV strain [22]. This has the theoretical advantage of avoiding continuous ventricular wall stress, potentially allowing longer periods of myocardial regeneration. In addition, a newly created atrial septal defect serves as a "pop-off" valve when afterload suddenly increases and the mLV is exposed to excessive (and potentially damaging) strain. An increased preload seems to promote the growth of the mLV, dynamically modulating the gradient across the PAB, providing intermittent training alternating with periods of relaxation to optimize myocardial regeneration [23].

Any patient with ccTGA is at risk of complete heart block, even without surgery, and it has been estimated to affect approximately 2% of patients per year after diagnosis. Surgically acquired heart block is often related to resection of the LVOTO or to closure of the VSD. The incidence of surgically acquired heart block is up to 20%, with freedom from pacemaker implantation being 70–80% at 15 years [4]. Regardless of cause, pacing has been strongly associated with mLV dysfunction [18].

Another important long-term concern is the development of sinus node dysfunction and atrial arrhythmias. Sinus node dysfunction has been reported in up to 40% of patients at 20 years of follow-up and atrial tachyarrhythmias in 20% of patients at 10 years of follow-up [24]. Available data suggest no difference between physiologic and anatomic repair with respect to the development of these problems [16]. Cavo-atrial isthmus-dependent atrial flutter is common and difficult to treat. This is the basis for considering prophylactic ablation as mentioned previously [6,23].

SUMMARY

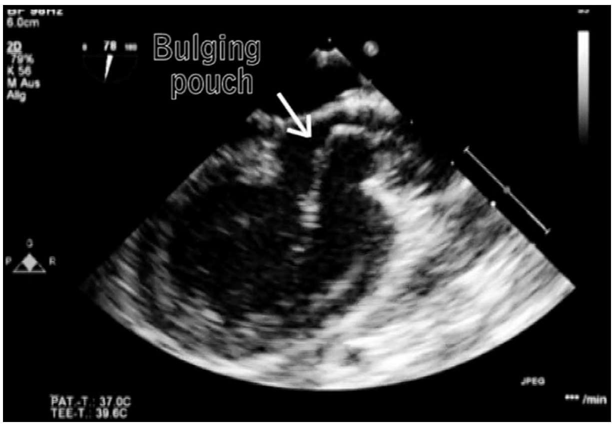
While we have gained considerable understanding, we do not yet fully know the optimal management strategy for every patient with ccTGA. As we are lacking in large systematic studies, it is challenging to provide well-grounded data-driven recommendations. We offer the following summary points:

1. it is obvious that leaving the mRV as the systemic ventricle runs counter to the wisdom of nature's design and will lead to premature heart failure for the majority of patients, albeit with many living to the fifth or sixth decade of life before heart failure ensues;
2. long-term survival and functional benefits after anatomic correction have been well demonstrated in the group of patients with preoperative TV regurgitation and mRV dysfunction;
3. the optimal method(s) and upper age limit of mLV training require further understanding;
4. the best outcomes with DS are achieved with patients in the first few years of life—even if reconditioning of the mLV is required, recent outcomes with anatomic repair in selected centers are very encouraging.

Careful selection of patients is needed, with some patients better served by other surgical options. Despite the challenges and the unknowns, we believe that it is in the best interest of the patients and the field to further refine our surgical approaches utilizing the mLV as the systemic pumping chamber and the MV as the systemic AV valve in ccTGA. In our opinion, these efforts to further refine care and provide consistent, excellent results should occur in centers with established expertise in managing patients with ccTGA [23].

SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:



Video 1 Double switch operation for corrected transposition of the great arteries {S,L,L} with resection of the subpulmonary obstruction in situs solitus (From: Hraska V, Murin P. Surgical management of congenital heart disease I: complex transposition of great arteries right and left ventricular outflow tract obstruction, Ebstein's Anomaly. A Video Manual. Heidelberg: Springer; 2012. Permission granted).

REFERENCES

[1] Dobson R, Danton M, Nicola W, et al: The natural and unnatural history of the systemic right ventricle in adult survivors. J Thorac Cardiovasc Surg 2013;145:1493–1501

[2] Graham Jr TP, Bernard YD, Mellen BG, et al: Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. J Am Coll Cardiol 2000;36:255–261

[3] Ilbawi MN, DeLeon SY, Backer CL, et al: An alternative approach to the surgical management of physiologically corrected transposition which ventricular septal defect and pulmonary stenosis or atresia. J Thorac Cardiovasc Surg 1990;100:410–415

[4] Hraska V, Vergnat M, Zartner P, et al: Promising outcome of anatomical correction of corrected transposition of the great arteries. Ann Thorac Surg 2017;104:650–656

[5] Zartner PA, Schneider MB, Asfour B, et al: Enhanced left ventricular training in corrected transposition of the great arteries by increasing the preload. Eur J Cardiothorac Surg 2015. https://doi.org/10.1093/ejcts/ezv416

[6] Hofferberth SC, Alexander ME, Mah DY, et al: Impact of pacing on systemic ventricular function in L-transposition of the great arteries. J Thorac Cardiovasc Surg 2016;151:131–138

[7] Chan DP, Van Hare GF, Mackall JA, et al: Importance of atrial flutter isthmus in postoperative intra-atrial reentrant tachycardia. Circulation 2000;102:1283–1290

[8] Murtuza B, Barron DJ, Stumper O, et al: Anatomic repair for congenitally corrected transposition of the great arteries: a single-institution 19-year experience. J Thorac Cardiovasc Surg 2011;142:1348–1357

[9] Hraska V, Mattes A, Haun C, et al: Functional outcome of anatomical correction of the great arteries. Eur J Cardiothorac Surg 2011;40:1227–1234

[10] El-Zein C, Subramanian S, Ilbawi M: Evolution of the surgical approach to congenitally corrected transposition of the great arteries. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2015;18:25–33

[11] Shin'oka T, Kurosawa H, Imai Y, et al: Outcomes of definitive surgical repair for congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections: risk analyses in 189 patients. J Thorac Cardiovasc Surg 2007;133:1318–1328

[12] Hraska V, Duncan BW, Mayer JE, et al: Long term outcome of surgically treated patients with corrected transposition of the great arteries. J Thorac Cardiovasc Surg 2005;129:182–191

[13] Lim HG, Lee JR, Kim YJ, et al: Outcomes of biventricular repair for congenitally corrected transposition of the great arteries. Ann Thorac Surg 2010;90:159–167

[14] DeLeon LE, Mery CM, Verm RA, et al: Mid-term outcomes in patients with congenitally corrected transposition of the great arteries: a single center experience. J Am Coll Surg 2017;224:707–715

[15] Said SM, Dearani JA: Physiologic repair of congenitally corrected transposition of the great arteries. In: Lacour-Gayet F, Bove EL, Hraska V, Morell VO, Spray TL, eds. Surgery of Conotruncal Anomalies. Cham, Heidelberg, New York, Dordrecht, London: Springer; 2016:495–515

[16] Fredriksen PM, Chen A, Veldtman G, et al: Exercise capacity in adult patients with congenitally corrected transposition of the great arteries. Heart 2001;85:191–195

[17] Helsen F, De Meester P, Van Keer J, et al: Pulmonary outflow obstruction protects against heart failure in adults with congenitally corrected transposition of the great arteries. Int J Cardiol 2015;196:1–6

[18] Baustista-Hernandez V, Marx GR, Gauvreau K, et al: Determinants of left ventricular dysfunction after anatomic repair of congenitally corrected transposition of the great arteries. Ann Thorac Surg 2006;82:2059–2065

[19] Barron DJ: Corrected TGA-VSD: the double switch procedure. In: Lacour-Gayet F, Bove EL, Hraska V, Morell VO, Spray TL, eds. Surgery of Conotruncal Anomalies. Cham, Heidelberg, New York, Dordrecht, London: Springer; 2016:441–458

[20] Myers PO, Del Nido PJ, Geva T, et al: Impact of age and duration of banding on left ventricular preparation before anatomic repair for congenitally corrected transposition of the great arteries. Ann Thorac Surg 2013;96:603–610

[21] Webb G, Gatzoulis MA: Atrial septal defects in the adult: recent progress and overview. Circulation 2006;114:1645–1653

[22] Van De Bruaene A, De MP, Buys R, et al: Right ventricular load and function during exercise in patients with open and closed atrial septal defect type secundum. Eur J Prev Cardiol 2013;20:597–604

[23] Hraska V, Mitchell ME, Woods RK, et al: What surgical improvements are needed to prove that anatomical repair is superior to physiological repair in the majority of patients with ccTGA. J Thorac Cardiovasc Surg 2017;154:1019–1022

[24] Dos L, Teruel L, Ferreria IJ, et al: Late outcome of Senning and Mustard procedures for correction of transposition of the great arteries. Heart 2005;91:652–656