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# Repaired Complete Transposition of the Great Arteries

# 10

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## Abstract

Complete transposition of the great arteries (d-TGA) is a common congenital heart disease. The surgical strategy for d-TGA has changed from the atrial switch operation to the arterial switch operation. This chapter describes the postoperative concerns regarding these two switch operations as well as details of the management of adult patients with repaired d-TGA. Long-term complications after the atrial switch operation include intra-atrial baffle-related problems, arrhythmias, right ventricular (systemic ventricular) failure, and tricuspid regurgitation. Transcatheter reintervention for baffle leaks or obstructions has been reported as effective. Right ventricular failure is common, and reintervention options for this are cardiac resynchronization therapy, arterial switch conversion, and cardiac transplantation. Atrial arrhythmias after the atrial switch operation are frequent. Catheter ablation and/or pacemaker implantation is indicated for some patients; however, the approach is complex. In recent years, the arterial switch operation has been the surgical option of choice. This procedure has the advantage of maintaining an almost normal anatomy. Long-term sequelae after the arterial switch procedure include pulmonary artery stenosis, coronary artery complications, and aortic valvular dysfunction. When pulmonary artery stenosis is severe, surgical repair should be performed. Sequential coronary angiography can reveal coronary stenoses in asymptomatic patients. The fate of the neo-aortic valve has been a major concern. In conclusion, there remain a number of concerns regarding repaired d-TGA patients; regular follow-up and appropriate timing of reinterventions are important.

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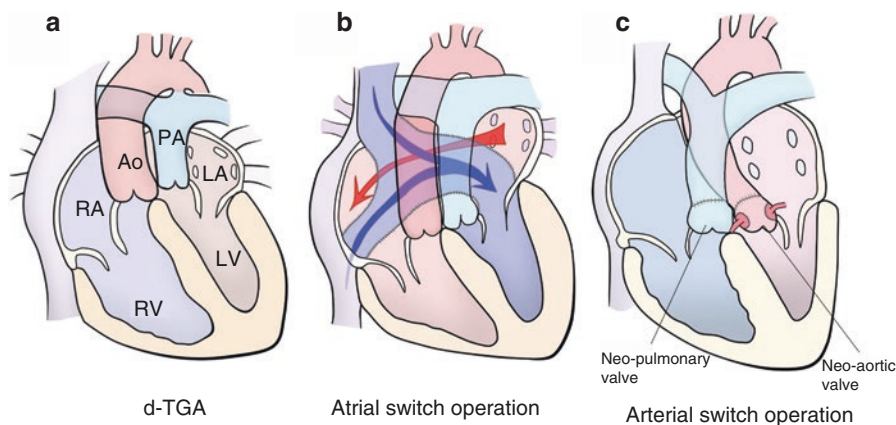
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### Keywords

Complete transposition of the great arteries • Adult congenital heart disease • Atrial switch operation • Arterial switch operation • Long-term complications

## 10.1 Introduction

Complete transposition of the great arteries (d-TGA; the “d” refers to the dextroposition of the bulboventricular loop) is defined as atrioventricular concordance and ventriculoarterial discordance and is characterized by the aorta arising from the morphological right ventricle and the main pulmonary artery arising from the morphological left ventricle (Fig. 10.1a). It is the second most common congenital heart disease that results in cyanosis (5–7% of congenital heart disease). Senning [1] reported the first successful case of the systemic and pulmonary venous switching using a right atrial wall and an atrial septum in 1959. In 1964, Mustard [2] reported a new procedure of systemic and pulmonary venous switching using a pericardial baffle in an atrium. Between the 1960s and mid-1980s, the physiological type of repair (Fig. 10.1b), which uses a venous switching technique at an atrial level, was the surgical treatment of choice. In patients with this repair, however, the morphological right ventricle functions as a systemic ventricle. In 1975, Jatene et al. [3] reported the first successful use of the arterial switch operation (Fig. 10.1c). Since then, several technical modifications, along with surgeons’ experience, have resulted in the standardization of this operation. Currently, the arterial switch procedure is the standard care for d-TGA in neonates.



**Fig. 10.1** (a) Schema in a patient with d-TGA. (b) After the atrial switch operation, blood from the superior and inferior vena cava was redirected to the mitral valve and pulmonary venous blood to the tricuspid valve in the atrium. (c) The arterial switch operation is often referred to as an anatomical correction, giving almost normal hemodynamics to a patient with d-TGA. RA right atrium, RV right ventricle, Ao aorta, PA pulmonary artery, LA left atrium, LV left ventricle

This chapter discusses the postoperative concerns regarding these operations and the management of adults with repaired d-TGA. Details of the Rastelli procedure and other procedures for d-TGA with left ventricular outflow tract obstruction are described in Chap. 9.

## 10.2 Atrial Switch Operation

Since the mid-1980s, atrial switch procedures have been superseded by arterial switch procedures, which means that patients with Mustard or Senning repairs will disappear over the next 40 years [4]. In recent years, the atrial switch operation has been adopted only as part of the double switch operation for corrected transposition of the great arteries. Many patients who underwent the atrial switch operation survived to adulthood, even beyond 40 years of age. The survival rates of the Senning and Mustard procedures have been reported as similar. Long-term complications include intra-atrial baffle-related problems, arrhythmias, right ventricular (systemic ventricular) failure, and tricuspid regurgitation (TR) [5, 6]. Indications for reintervention after the atrial switch operation are summarized in Table 10.1.

### 10.2.1 Intra-atrial Baffle-Related Complications

Intra-atrial baffle-related problems include baffle leaks and obstructions. Khairy et al. [7] reported that baffle-related complications occur in as many as one in five adult survivors. These complications account for majority of the reoperations, but the mortality risk of reoperations has been reported to be high, at 36% [8]. Recently, catheter techniques for treating baffle leaks and obstructions have been reported.

**Table 10.1** Indications for reintervention after the atrial switch operation

Complications	Indications	Procedures
Baffle leaks	<ul style="list-style-type: none"> <li>• Large shunt</li> <li>• Tachyarrhythmias</li> <li>• Presence of endocardial pacemaker</li> </ul>	<ul style="list-style-type: none"> <li>• Surgery</li> <li>• Transcatheter procedures</li> </ul>
Baffle obstruction	<ul style="list-style-type: none"> <li>• High pressure gradient (10–15 mmHg)</li> <li>• Symptoms (SVC syndrome, PLE)</li> <li>• Pacemaker implantation</li> </ul>	<ul style="list-style-type: none"> <li>• Surgery</li> <li>• Transcatheter procedures</li> </ul>
Systemic right ventricular failure	<ul style="list-style-type: none"> <li>• Resistant to medical therapy</li> </ul>	<ul style="list-style-type: none"> <li>• CRT</li> <li>• Retraining of left ventricle</li> <li>• Transplantation</li> </ul>
Tricuspid valve regurgitation	<ul style="list-style-type: none"> <li>• Severe regurgitation</li> </ul>	<ul style="list-style-type: none"> <li>• Valve repair</li> <li>• Valve replacement</li> <li>• Pulmonary artery banding</li> </ul>

*SVC* superior vena cava, *PLE* protein-losing enteropathy, *CRT* cardiac resynchronization therapy

### 10.2.1.1 Baffle Leaks

Intra-atrial baffle leaks result in left-to-right shunts such as a patent fossa ovalis. Most of the leaks are small but multiple and are often visualized only by transesophageal echocardiography. Baffle leaks are best imaged using agitated saline injection performed during a transesophageal echocardiogram [9]. Three-dimensional transesophageal echocardiography can provide practical information regarding orientation in patients with complex anatomy [10].

A large shunt is an absolute indication for a transcatheter or surgical closure [11–13]. Even in patients with small baffle leaks, the risk of paradoxical embolus and cerebrovascular events may increase if the patient has tachyarrhythmias or an endocardial pacemaker [11].

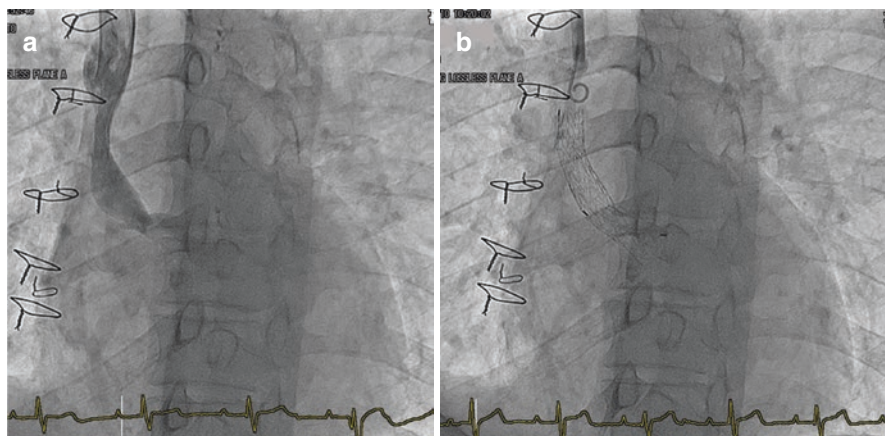
Although surgical repair of the atrial baffle has previously been reported, catheter interventions may be safer and more effective [12, 14]. An atrial septal occluder is usually used for the closure of baffle leaks [12, 15–17]. Daehnert et al. [15] described the widely varying positions of the baffle leaks, which result in atypical occluder positions compared to native atrial septal defects. Precise position assessment using transesophageal echocardiogram in conjunction with fluoroscopy is important.

Using a covered stent has been reported as a method for simultaneously eliminating baffle leaks and obstructions [18, 19]. Recently, stent-graft devices (such as the Gore excluder<sup>®</sup> device) have been used; these are commercially available for use in patients with abdominal aortic aneurysm [20, 21].

### 10.2.2 Baffle Obstruction

Obstruction of the systemic venous pathway is one of the most common complications following the atrial switch operation. The superior limb is most commonly affected at 37–44% of the cases [11, 22]. Obstruction of the superior vena cava can result in chylothorax [23] and superior vena cava syndrome [24]. Inferior baffle obstruction may cause protein-losing enteropathy [25–27]. Although pulmonary vein obstruction is less common, it can occur both early and late in the postoperative period [28]. These obstructions can be imaged by transthoracic echocardiography [29]. The baffle obstruction should be assessed by transesophageal echocardiography [30], angiography, computed tomography (CT), or magnetic resonance imaging (MRI) to visualize anatomical details.

Although reintervention should be considered in symptomatic patients, asymptomatic patients for whom future pacemaker placement is planned should also be treated to prevent further stenosis of the venous pathway. Indications for reintervention for venous stenosis are still under debate. Some reported cases have suggested that a high pressure gradient (>10–15 mmHg) or the presence of symptoms (e.g., superior vena cava syndrome or protein-losing enteropathy) are indications [26, 27]. Insignificant baffle obstruction in asymptomatic patients with normal cardiac function can also be a risk factor for sudden death [31].



**Fig. 10.2** (a) Angiography showing stenoses of the superior vena cava and the baffle. (b) A Gore excluder® device was implanted

Although surgical reintervention methods have been reported [13, 32], catheter techniques have also been introduced for this complication. Balloon angioplasty has demonstrated effectiveness in the short term but has a high incidence of restenosis of up to 40% [33]. Stent implantation is therefore recommended [34, 35]. As patients with baffle obstruction frequently have concomitant baffle leaks, a covered stent can be used as simultaneous treatment for both the baffle obstructions and leaks [20, 21]. Figure 10.2 shows an angiogram of the placement of a Gore excluder® device for the superior vena cava and baffle stenoses.

Laser lead extraction is necessary before the transcatheter procedure in patients with an atrioventricular block and endocardial pacemaker [21].

### 10.2.3 Systemic Right Ventricular Failure

Morphological right ventricular failure has been recognized in the long term after the atrial switch operation [36]. The mechanism for this is not well understood. One possible mechanism results from the structural difference between the left and right ventricles [37]. The left ventricle has a limited nontrabeculated area and two papillary muscles, whereas the right ventricle has an infundibulum without significant trabeculation. The longitudinal arrangement of cardiomyocytes in the right ventricle may be a morphological cause of the inability to compensate for high volume and pressure, and myocardial fibrosis [5], myocardial ischemia [38], and ventricular–ventricular interaction [39] have been described as reasons. The association of TR and/or arrhythmias can lead to progressively deteriorating ventricular dysfunction.

MRI and echocardiography are useful tools for evaluating right ventricular function. Although cardiac MRI is the gold standard for this assessment [40], it cannot

be used in patients with pacemakers. Echocardiography is a more commonly used modality, but its usefulness of evaluation on right ventricle is limited by the shape of right ventricle and the expertise of the echocardiographer. The myocardial performance index has been used to evaluate heart failure after an atrial switch procedure [41]. More recently, Diller et al. [42] reported that a reduction in global longitudinal strain is a good predictor of adverse clinical outcome in these patients.

Medical management using angiotensin-converting enzyme inhibitors [43, 44], angiotensin receptor blockers [45], aldosterone antagonists [46], and beta-adrenergic receptor blockers [47] has been reported and discussed, but left ventricular failure management principles cannot be applied to right ventricular failure [44]. When these medical management methods are not effective, heart failure should be treated by reintervention. Options for this include cardiac resynchronization therapy (CRT), arterial switch conversion with retraining of the left ventricle (i.e., pulmonary artery banding), and cardiac transplantation.

CRT using biventricular pacing has been shown to be beneficial in patients with left ventricular failure, whereas the efficacy of CRT in patients with right ventricular failure has not yet been established. Particularly for patients with associated congenital heart disease, the benefit of CRT is still unclear [48]. Although there have been no large clinical studies, some promising reports have described CRT as improving hemodynamics in adult congenital heart disease with a systemic right ventricle [49, 50]. If the coronary sinus is accessible from the systemic veins, the systemic right ventricular lead may be placed entirely transvenously [51]; the alternative would be the implantation of epicardial leads with thoracotomy. In our institution, two patients with Mustard repair underwent the replacement of a CRT device, and in one of them we took a hybrid approach involving the implantation of leads. After the right atrium and left ventricular leads had been placed percutaneously, the right ventricular lead was placed with thoracotomy. Pacing site selection and setting the CRT (e.g., A-V delay, V-V delay) remain complex. There has been a reported case of a patient with Mustard repair who underwent upgrading from a single-chamber pacemaker to CRT, and functional evaluation revealed improved cardiac performance in DDD mode with broad QRS [52]. This result seems to indicate that the data derived from CRT in structurally normal left ventricular failure are not reliable for the treatment of right ventricular failure with congenital heart disease.

Retraining of the left ventricle and conversion to the arterial switch protocol was introduced by Mee in 1986 [53]. Some reports have described successful outcomes in young patients [54]. Poirier et al. [55] reported that being >12 years old at the time of the operation was a risk factor for mortality. In some adult cases, the left ventricle cannot respond to the increased afterload, and then biventricular failure occurs. In general, retraining of the left ventricle for conversion to the arterial switch operation cannot be an option in adults. Conversely, the increased afterload may result in a rightward “septal shift” and an improvement of tricuspid valve coaptation. The regression of TR after pulmonary artery banding has been observed in some patients [56, 57]. A similar mechanism may explain the superior prognosis in patients with congenitally corrected transposition of the great arteries (ccTGA) with pulmonary stenosis

compared with ccTGA without pulmonary stenosis [58]. Whether pulmonary artery banding provides effective palliation has yet to be determined.

Orthotopic heart transplantation should be considered for patients with systemic right ventricular failure not amenable to medication or the surgical options described above. Irving et al. [59] described their experience of cardiac transplantations in the adult population with congenital heart disease and reported that 22% of transplants were performed in d-TGA following atrial switch. In the same report, 88% in the d-TGA patients survived at the early period of the cardiac transplantation, and only one death at 7 years was described.

### 10.2.4 Tricuspid Valve Regurgitation

Mild TR is very common in adult patients who underwent Mustard or Senning procedures. Moderate-to-severe TR is found in more than 20% of the adult survivors [60]. Etiologies of these TRs include structural abnormalities of the valve itself, iatrogenic lesions during the Rashkind balloon atrial septostomy [61] or prior ventricular septal defect repair, and functional regurgitations secondary to the right ventricular dysfunction. In d-TGA patients, intrinsic abnormality of the tricuspid valve is very rare [62, 63]. Szymański et al. [60] analyzed 42 patients with d-TGA after the atrial switch operation and classified their TRs into the following three distinct types based on their causes: predominant annular dilatation (type I), valve prolapse (type II), and valve tethering (type III). These authors reported that the most common type was tricuspid valve tethering (44%). In addition to a mechanism similar to functional mitral regurgitation, a right-to-left septal shift may contribute to the tethering. Beta-adrenergic receptor blockers have been suggested as medications for functional TR [64]. If TR becomes severe, tricuspid valve repair or replacement may be required.

The surgical option, i.e., replacement or repair of the tricuspid valve, is still controversial [65]. This valve can be approached via the right atrium. Tricuspid valvuloplasty is often performed with the technique of annuloplasty using a ring. The recurrence rate of TR after these repairs is reported to be up to 37% [65]. Although tricuspid replacement is required either when the quality of the leaflets is insufficient or when recurrence occurs, the results can be disappointing, with little improvement in hemodynamics [66, 67]. Pulmonary artery banding may be a better option as previously described, although currently there is insufficient data demonstrating favorable survival after this procedure. Patients with symptomatic right ventricular failure associated with TR may ultimately require cardiac transplantation.

### 10.2.5 Arrhythmias

Long-term bradyarrhythmias and tachyarrhythmias are common in patients after the atrial switch operation [68]. Bradyarrhythmias are generally induced by the sick sinus syndrome (SSS); Gellat et al. [69] reported that SSS was found in 23% at

5 years and 60% at 20 years of survivors following the atrial switch operation. These may be caused by damage during the initial operation or the interruption of right atrial blood flow. Pacemaker implantation is required when SSS occurs; however, some considerations around pacemaker implantation need to be taken into account. In the presence of an intra-atrial baffle leak, the risk of systemic thromboemboli will be more than twofold with transvenous leads implantation [70]. The patency of the baffle route should be assessed before implantation. In patients who have stenosis of the superior vena cava or the superior baffle, a catheter intervention before implantation or epicardial pacing may be necessary. Finally, consideration of the potential benefit of CRT is important for the prevention of subsequent right ventricular failure.

Atrial tachyarrhythmias are found in up to 25% of patients, and intra-atrial reentrant tachycardia (IART), also referred to as atrial flutter, is the most frequent type of arrhythmia [7, 69]. IART has been reported to be a probable marker for sudden death [71], and atrial arrhythmias may be a surrogate marker for ventricular dysfunction [72]. Pacemaker implantation may be considered not only in the setting of bradyarrhythmias but also of tachyarrhythmias to facilitate aggressive medical therapy.

Catheter ablation is indicated for patients with recurrent symptomatic or drug refractory atrial arrhythmias. Traditional ablation procedures are limited by the anatomic complexity in these patients. Most IART circuits are similar to typical atrial flutter in a heart with a normal structure, but the cavotricuspid isthmus is partitioned into two distinct regions by the atrial baffle. The tricuspid valve is not directly accessible from the systemic venous route because of this partition; therefore, another approach is required such as access via a baffle leak, a transbaffle puncture [73], or a retrograde aortic approach using remote magnetic navigation [74]. A transbaffle puncture under transesophageal echocardiographic guidance is safe and effective [73, 75]. El-Said et al. [76] reported their 18-year experience with 16 patients who had previously undergone the atrial switch operation and suggested that transbaffle puncture was a safe technique with no residual shunt at follow-up. Although good results of a retrograde approach have also been reported [74], this brings the potential risk of damage to the atrioventricular valve, and it is impossible in the presence of a mechanical valve. Magnetic navigation technology is still only available in a few institutions in Japan, but the reduction in radiation exposure with this technique is particularly important.

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### 10.3 Arterial Switch Operation

Since the first introduction of the arterial switch operation for d-TGA by Jatene in 1975 [3], techniques and management for the care of neonatal congenital heart disease have improved. In 1984, Castaneda et al. [77] reported successful results in newborns with d-TGA and demonstrated the capacity of the left ventricle in a neonate to take over the systemic circulation. The transition in surgical strategy from



atrial switch to arterial switch procedures was mostly completed by the early 1990s. These days, it is usually performed in the first month of life for all forms of d-TGA without left ventricular outflow tract obstruction.

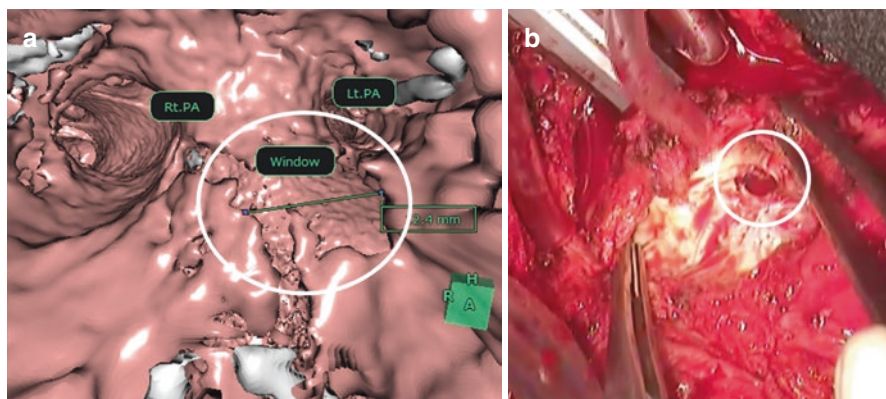
Losay et al. [78] reported one of the largest studies of arterial switch procedures, which found that the survival rate at 10 and 15 years was 88% and that there were no deaths after 5 years in the 1095 survivors. Because the perioperative outcome of the arterial switch operation has been reported to be good [79–81], the focus has shifted from mortality to long-term complications and exercise performance [82, 83]. The arterial switch procedure has the advantage of maintaining an almost normal anatomy and avoiding incisions and suture lines in the atrium. However, the technique involves translocating the aorta and the pulmonary artery and detachment of the coronary arteries in the neo-aorta. It is a delicate operation, and long-term sequelae after the procedure include pulmonary artery stenosis, coronary artery complications, and aortic valvular dysfunction. These complications may require surgical or transcatheter reintervention.

### 10.3.1 Supravalvular Pulmonary Artery Stenosis

Pulmonary artery stenosis is the most commonly encountered long-term complication in patients after the arterial switch operation [81, 84, 85], with the incidence reported to range from 3% to 30% [86]. Various causes have been reported, with the technique for the reconstruction of the pulmonary artery possibly being one of the most important factors. Although the direct anastomosis technique reported by Pacifico et al. [87] has the advantage of reconstruction without any artificial materials, the tension at the direct anastomotic site is higher than that for anastomosis with a patch [85]. After the Lecompte maneuver [88], the left pulmonary artery is likely to be narrow. Presence of the ductal tissue has also been suggested to be a cause of pulmonary artery stenosis [86].

Echocardiography, angiography, and CT are used to assess the degree of pulmonary artery stenosis. Catheterization should be performed to measure the right ventricular pressure. Patients with symptoms, severe stenosis of the branch pulmonary artery, or right ventricular systolic pressure >50 mmHg may require reintervention [5].

The reintervention-free rate for right outflow obstruction reported by Williams et al. [89] was 83% at 10 years. In the same report, 55% of 62 reinterventions were surgical, and the remaining 45% were percutaneous. Catheter intervention techniques, such as balloon angioplasty and stent implantation, have been reported for dilating branch pulmonary artery stenosis [90, 91]. Torres et al. [92] described 18 cases of iatrogenic aortopulmonary communication after catheter interventions, with nine patients requiring surgical closure of the communication. An inner vascular image by CT and an intraoperative photograph are shown in Fig. 10.3. When the obstruction is severe, which leads to a high pressure gradient, surgical repair is more clinically efficacious than a percutaneous procedure [80].



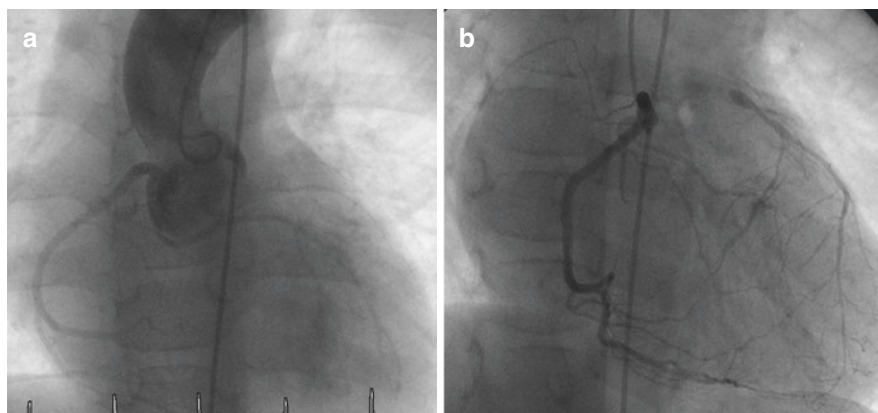
**Fig. 10.3** (a) An inner vascular image by CT shows a view of aorticopulmonary communication from the pulmonary artery. (b) An intraoperative photograph; the white circle indicates a window to the aorta. The aorticopulmonary communication was repaired with a heterologous pericardium patch. *Rt.PA* right pulmonary artery, *Lt.PA* left pulmonary artery, *Window* aorticopulmonary communication

### 10.3.2 Coronary Artery Stenoses or Obstructions

In the arterial switch procedure, reimplantations of coronary arteries should be performed; this is not required in the atrial switch procedure. Some problems related to replaced coronary arteries may occur both early and long term postoperatively. Perioperative coronary complications have a significant impact on the results of the operation [93–95]. A meta-analysis by Pasquali et al. [96] revealed that intramural or single coronary artery patterns were significant risk factors for operative mortality. The incidence of late coronary artery stenosis or obstruction after the arterial switch operation has been reported to be 3–7% [97–100]. Although coronary artery stenosis or obstruction has been reported to be a risk factor for sudden death [97, 101], most patients with late coronary artery stenosis are asymptomatic. It is possible even for the total obstruction of the coronary artery to be found in asymptomatic patients [98]. The richness of retrograde coronary perfusion from collateral arteries or cardiac denervation [102] is one of the mechanisms for this asymptomaticity.

Anatomical kinking, extrinsic compression, stretching, and intimal proliferation are possible causes of reimplanted coronary stenosis or obstruction [98]. Studies in animals [103] and humans [104] have described the normal growth of aorta–coronary anastomoses after the arterial switch operation. Intracoronary ultrasound assessment has revealed proximal intimal thickening in 89% of patients in the long term following the arterial switch operation [105], and this change may be related to the occurrence of late coronary ostial stenosis.

Noninvasive modalities, such as electrocardiogram, echocardiography, and exercise testing, have low sensitivity for coronary stenosis after the arterial switch operation [99, 106]. Although selective coronary angiography is the most accurate means for assessing the patency of reimplanted coronary arteries, the indication for



**Fig. 10.4** (a) Coronary angiography in a d-TGA patient after the arterial switch operation. The reimplanted right coronary artery and circumflex artery were patent, and the ostium of the left descending artery was totally occluded. (b) The left descending artery was found with flow of collateral arteries from the right coronary artery and circumflex artery. This patient was asymptomatic and was treated with a coronary artery bypass graft

this invasive examination has not been determined because most patients with coronary stenosis or obstruction after the arterial switch operation are asymptomatic as previously mentioned. Angiography for an asymptomatic patient with ostial occlusion of a reimplanted left descending artery is shown in Fig. 10.4. Legendre et al. [99] recommended sequential selective coronary angiography at 5, 10, and 15 years following the operation. However, noninvasive techniques for the assessment of coronary stenosis have been developing. Multislice CT is one alternative modality for detecting ostial stenosis in patients after the arterial switch operation [107]. Coronary assessment using MRI has an advantage over other non-X-ray techniques; however its diagnostic accuracy remains unsatisfactory because of its low spatial resolution [108]. Scintigraphy with exercise or dynamic positron emission tomography with N-13 ammonia during pharmacologic vasodilation is used to assess the physiological significance of coronary artery stenosis [109]. Even in asymptomatic patients with good hemodynamic condition, a continuous follow-up of the coronary arteries after the arterial switch operation is of major importance. In patients with severe coronary stenosis or obstruction, treatment should be undertaken to prevent sudden death.

Surgical or transcatheter procedures are options for the treatment of coronary stenoses. Coronary artery bypass grafting (CABG) and ostial patch angioplasty are surgical options. It is important to select the appropriate type of revascularization procedure to be performed in each case [110]. In most reported cases with CABG, internal thoracic artery (ITA) was used for the bypass graft [111, 112], and excellent midterm results have been reported after CABG with ITA in a young population [113]. CABG with bilateral ITAs has also been reported [110, 114]. ITAs are live grafts with growth, and their long-term patency is expected. The other surgical option is ostial patch plasty of the coronary artery. Segmental stenosis of a coronary artery

ostium in the absence of calcification is indicated for this procedure. An autologous pericardium has been used in many cases to enlarge ostial stenosis [115]. A saphenous vein patch or polytetrafluoroethylene patch has also been used in some cases [116]. A simple ostial stenosis can be repaired by onlay patch enlargement from the main coronary stem to the adjacent aortic wall. In a patient with a single coronary ostium, modified coronary ostial angioplasty that employed a single, “pantaloon”-shaped autologous pericardial patch has been reported [110]. The coronary artery pattern and the relationship between the aorta and the pulmonary artery should be assessed preoperatively. To allow adequate exposure of the aortic root, the pulmonary trunk should be transected under some anatomical conditions. Percutaneous balloon dilation of the ostial stenosis is not effective [117]. Although percutaneous stenting may be effective, implantation of a stent in the ostial position of a coronary artery or in a young population can be potentially harmful. The sequential redilation of stents may be needed, and in patients with stent implantation in the coronary ostium, a subsequent surgical ostial patch plasty may be difficult or impossible. The redilation of stents in congenital heart disease is reported to be safe and effective [118]. Both bare-metal stents and drug-eluting stents can be used for the implantation. An antithrombosis prophylaxis is essential after stenting.

### 10.3.3 Neo-aortic Valve Insufficiency

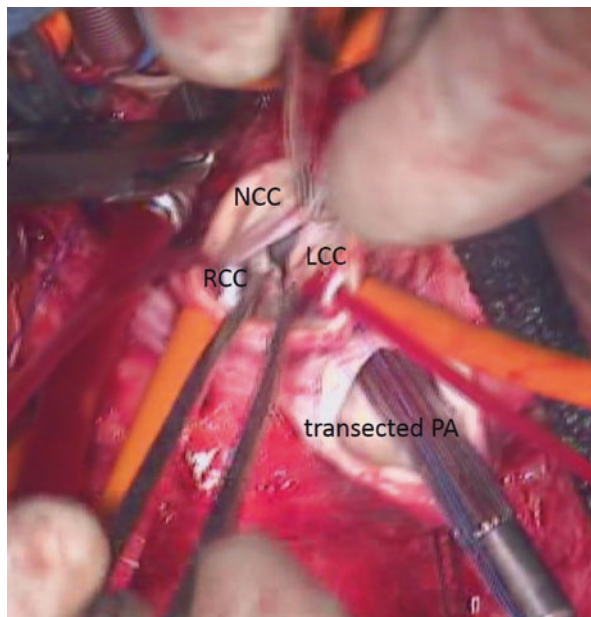
The fate of the neo-aortic valve, which is the native pulmonary valve, after the arterial switch operation has been a major concern since the operation was introduced. Neo-aortic valve insufficiency has been reported with prevalences varying between 0.3% and 55% [78, 84, 119–121]. Although the incidence of reintervention for neo-aortic valve insufficiency has been reported to be low [111, 122], the insufficiency has been described as following a time-dependent progression [123]. Reinterventions for neo-aortic insufficiency may become more frequent in the near future.

The etiology of neo-aortic insufficiency has been reported to be multifactorial [118, 123]. Possible risk factors include prior pulmonary artery banding (two-stage anatomical repair), the presence of a left ventricular outflow tract obstruction [124], the presence of a ventricular septal defect [122, 123], and the trapdoor type of coronary reimplantation [121]. It has been shown that the pulmonary valvular leaflets are able to mechanically tolerate the systemic pressure in patients following the Ross procedure. Neo-aortic dilatation is one of the associated concerns; however, some authors have reported no correlation between neo-aortic insufficiency and root dilatation [121, 124, 125].

Neo-aortic valve insufficiency can easily be found by transthoracic echocardiography [119]. Patients with severe incompetence should be referred for surgical reintervention before left ventricular dysfunction occurs [123].

Suggested surgical treatment options for neo-aortic insufficiency include various repair techniques [126], neo-aortic valve replacement [127], and the Ross operation [128]. In the young population, valve replacement should be avoided. Serraf et al. [111] reported the application of various techniques, such as annuloplasty [129] and

**Fig. 10.5** Neo-aortic cusps were inspected after the main pulmonary trunk was transected. The right coronary cusp was dysplastic. The cusp was enlarged with an autologous pericardial patch. *NCC* noncoronary cusp, *RCC* right coronary cusp, *LCC* left coronary cusp, *PA* pulmonary artery



commissuroplasty [130], to neo-aortic valve repair. Transection of the main pulmonary trunk may be necessary to approach the neo-aortic root. Figure 10.5 shows an intraoperative photograph of one of our cases of neo-aortic valve repair after the arterial switch operation. When the neo-aortic root dilatation is concomitant, the Bentall operation or valve-sparing reimplantation technique [131] should be employed.

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