Late coronary complications after arterial switch operation for transposition of great arteries. Clinical and therapeutic implications.

Abstract

The arterial switch operation is the procedure of choice for correction of transposition of the great arteries. Although offering excellent long term results, this procedure is burdened by the risk of acute and subacute coronary occlusion. No guide-lines exist for the management of acute or chronic ischemia in this setting. We briefly review the literature and present the results of our institution.

MeSH: Transposition of Great Vessels, Coronary Vessels, Heart Defects, Congenital/surgery, Diagnostic Imaging, Coronary Angiography

Introduction
The arterial switch operation (ASO) has become the procedure of choice for correction of transposition of great arteries (TGA). Compared with the Mustard and Senning repairs, correction at the arterial level restores the left ventricle as the systemic ventricle and offers improved outcome. Coronary artery transfer was first described by Wernovsky in 1993 (figures 1,2).¹

Figure 1 Coronary arteries transfer

![Figure 1](image1)

Figure 2 Coronary anatomy in transposition of the great arteries in accordance to Wernovsky

![Figure 2](image2)

The transfer of the coronary arteries during ASO may be a difficult step in the case of abnormality of origin or distribution of these arteries and this technique is burdened by the risk of acute and subacute coronary occlusion.² Various classifications of coronary anatomy in transposition of the great arteries exist. In our department we define coronary anatomy in accordance with Yacoub’s classification (figure 2). Comparison of mortality between usual pattern and variant patterns showed that the presence of a single coronary and an intramural coronary was associated with a higher risk of occlusion (figure 3).²

No guidelines exist concerning the management of coronary artery obstructions after ASO. We recently reviewed our institutional experience concerning the early and long term outcome of ASO, in order to: determine the prevalence of coronary arteries obstruction, establish possible implications between obstruction and late death, try to

identify mechanisms involved in early and late coronary obstruction and propose a therapeutic approach. We present here our results.

Figure 3 Yacoub classification of coronary arteries anatomy in transposition of the great arteries. Type A: Left coronary artery takes origin from the left sinus and right coronary artery from the right sinus. Type B: Single coronary artery. Type C: Two para-commissural ostia with or without intramural course. Type D: RCA and circumflex take origin from the right ostium, LDA alone takes origin from the left ostium. Type E: RCA and LDA take origin from the left ostium, circumflex alone takes origin from the right ostium.
Patients and methods
The series comprises 712 patients who underwent ASO in the neonatal period for simple and complex transposition of the great arteries. All patients had clinical evaluation, ECG and echocardiography 6 months after surgery and every 6 months, until selective coronary angiography was performed. Coronary angiography was performed: 1) Systematically at the age of 5 years in asymptomatic patients, 2) When an ischemic event occurred, 3) When electrocardiographic and/or echocardiographic findings suggesting myocardial ischemia were detected. Myocardial perfusion imaging was performed: 1) Systematically at the age of 3 years in asymptomatic patients, 2) When an ischemic event occurred 3) 6 months after surgical revascularization, percutaneous or medical treatment of coronary obstruction. Myocardial ischemia was defined as the presence of ST-T changes/Q waves on ECG or ultrasound anomalies, together with perfusion defect at scintigraphy, consistent with the localization of coronary obstruction. Classification of late complication was: occlusion, major stenosis (>50% of coronary artery diameter), minor stenosis (< 50% of coronary artery diameter) or stretching (diffuse narrowing). Coronary diameter was measured at end-diastole. Functional status was determined on the basis of: clinical examination, electrocardiogram, echocardiography, selective coronary angiography, myocardial perfusion imaging (Thallium-201) at rest and after exercise or dipyridamole infusion.

Results
At a mean follow-up of 4.7 ± 2 years, we identified 39 children with coronary obstruction (5.5%). Thirty three patients had isolated TGA and 6 had TGA associated with ventricular septal defect and/or aortic coarctation. Mean age at diagnosis of coronary obstruction was 3.1 ± 3 years. Ten patients were identified from an anomalous selective coronary angiogram, performed during a prospective study, to determine the long term outcome of coronary anastomoses (figures 4,5).

Figure 4 Selective injection of the left ostium shows revascularization of the circumflex artery (LAO and RAO view)
Twenty nine patients were identified because of electrocardiographic and/or echocardiographic anomalies suggestive of myocardial ischemia. Two patients died suddenly at the age of 4 and 6 months. Both had a stenosis of the left ostium diagnosed one month after surgery. One patient died in the postoperative period of surgical revascularization. Type of coronary obstruction according with myocardial scintigraphy is illustrated in figure 6.

Sixteen patients (43%, median age 1.75 year) had a perfusion defect on first scintigraphy, consistent with the site of coronary obstruction. Only 3 of them had normal ECG and echocardiography. Twenty one (57%, median age 2 years) had normal first scintigraphy at rest and after exercise. Eight of them, however, had severe coronary lesions with collateral vessels. Eleven patients had normal ECG and echocardiography.

Fifteen patients received a surgical intervention, 4 had medical treatment and 17 had no treatment (figure 7). Our results and patients outcome are summarized in figure 8.

Figure 7 Therapeutical approach to coronary obstruction after ASO. Surgical treatment n= 15 (group 1), Medical treatment n=4 (group 2), No treatment n=17 (group 3), Death n=3 (group 4)

Figure 8 Flow chart summarising results and treatment of patients with coronary occlusion

Among 15 patients undergoing surgical revascularization (38.5%), 5 had a coronary artery bypass graft (CABG) by interposition of a segment of mammary artery, and 10 had a surgical angioplasty (figure 9), by interposition of a vein or subclavian patch. All children undergoing surgical treatment had elective coronary angiography 1 month after surgical revascularization. All coronary ostia were patent (figures 10-12). A discrepancy between the ostial region and the native coronary artery was noticed in most cases (figure 12). All patients undergoing surgical or medical treatment recovered normal perfusion at scintigraphy. Two patients who underwent PET showed a decreased coronary reserve. In 3 out of 17 patients with mild coronary obstruction and normal myocardial perfusion, who did not receive any treatment, a normal coronary angiography was found at follow-up.

Figure 9 Surgical left coronary angioplasty

Figure 10 Selective coronary angiography following surgical angioplasty by interposition of a venous patch (type D anatomy)
Figure 11 Selective right coronary angiography following surgical angioplasty by interposition of a veinous patch (type A anatomy)

Figure 12 Selective left coronary angiography following surgical angioplasty by interposition of a subclavian patch (type A anatomy)

Discussion
Arterial switch operation has dramatically changed the outcome of patients with TGA. Although initially burdened by an elevated mortality, this technique is now performed with good results in most centres. A previous report showed that in our department this operation has a 10 year survival of 94%, and a freedom from surgical reintervention of 78%.

Risk factors that have been demonstrated to influence early mortality are right ventricular hypoplasia, lower birth weight and longer intraoperative support.

Several authors underlined the occurrence of early or late coronary obstruction after ASO. The reported incidence of coronary obstruction varies between 5 and 8%. Some anatomical types are reported to have a higher incidence of early and/or late obstruction. In particular, in type B and type C anatomy, transfer of the coronary arteries may be a difficult step.

Echocardiography has been show to be able to detect the presence of coronary anomalies and in particular to unmask an intramural course. Preoperative diagnosis is advantageous, because it helps to prevent accidental injury to the intramural coronary artery during transaction of he aortic root and excision of the coronary
artery ostium from the aorta. In most centres, however, precise coronary anatomy is usually defined at surgery.

Diagnosis of ischemic obstruction is typically performed when an acute ischemic event occurs in the postoperative period or, after discharge, on the basis of echocardiographic signs of ischemia.

No guidelines exist concerning the follow up of children undergoing ASO. Different techniques have been reported to be a useful tool for diagnosis of coronary obstruction in this setting. In our department we decided to systematically perform coronary angiography and myocardial scintigraphy. We previously reported the usefulness of selective coronary angiography for detection of coronary obstruction in this setting. Even if invasive and potentially dangerous, selective coronary angiography allowed us to identify a subgroup of high-risk children with normal echocardiography. We also showed that aortic root angiography yields ambiguous results in ASO patients. In the absence of coronary lesions at systematic coronary angiography, children are monitored noninvasively after 2 and 5 years.

Myocardial perfusion imaging by thallium-201 is useful in selecting patients with coronary artery lesions who are candidates for myocardial revascularization. However, this technique does not identify hibernating territories that are expected to recover a normal contraction after revascularization. On the contrary, positron emission tomography (PET) assesses myocardial viability by coronary blood flow and metabolic myocardial activity. It allows the targeting of angioplasty to hibernating territories that have residual metabolic activity. It also allows the evaluation of coronary flow reserve. Other authors have underlined that ASO patients have a diminished flow reserve. The global impairment of stress flow dynamics may indicate altered vasoreactivity. Although noninvasive, this technique is irradiating and has a limited availability.

Prognostic significance of reduced coronary reserve in ASO patients needs to be determined. In particular, the long term impact on cardiac function has not been established. It has been demonstrated that aorto-coronary anastomoses can grow. However, alterations of sympathetic innervation after surgery is a well known phenomenon, that could justify an impaired regulation of coronary flow in ASO patients. It has also been shown that endothelial function could be altered in this setting. The prognostic significance of these findings needs to be determined.

Surgical angioplasty or medical treatment can be carried out to restore coronary perfusion. Detection of symptom free obstructions and their treatment are necessary for late myocardial protection. In fact, obstruction may progress and collateral circulation may become insufficient with growth. Treatment of coronary obstruction allows decreased side effects of chronic myocardial hibernation such as degeneration and ventricular dysfunction. Previous report have shown that early treatment can preserve myocardial function over long time. Surgical angioplasty of the coronary stems restores physiological coronary perfusion and patch enlargement of coronary ostia offers encouraging mid-term results. Internal thoracic or mammary artery grafting can be considered a valid alternative to surgical angioplasty in the presence of long stenosis.

Conclusions

Symptom free coronary obstruction is not infrequent after ASO. We recommend systematic selective coronary angiography for detection of coronary obstruction. Non-invasive methods to evaluate these patients will be hopefully available soon. Surgical revascularization should be performed to preserve myocardial function and avoid late ischemic events in case of myocardial ischemia. However, concern exists with regard to late outcome of coronary revascularization in children and on durability.
of coronary angioplasty. Optimal treatment of coronary obstruction in ASO patients is not consensual.

References


