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Total anomalous pulmonary venous connection: long-term appraisal with evolving technical solutions☆

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Abstract

Objectives: To evaluate late outcome of non-isomeric total anomalous pulmonary venous connection (TAPVC) repair, controlling for anatomic subtypes and surgical technique. **Methods**: Between 1983 and 2001, 89 patients (median age 54 days) underwent repair for supracardiac (38), cardiac (26), infracardiac (16) or mixed (nine) TAPVC. Ten patients (11.2%) presented associated anomalies other than PDA. Twenty-eight patients (31.5%) were emergencies, due to obstructed drainage. Supracardiac and infracardiac TAPVC repair included the double-patch technique with left atrial enlargement in 29 patients and side-to-side anastomosis between the pulmonary venous (PV) confluence and the left atrium in 29 patients. Coronary sinus unroofing was preferred for cardiac TAPVC repair. Total follow-up was 727.16 patient-years (mean 8.55 years, 98.8% complete). **Results**: Early mortality was 7.86% (7/89). Ten patients (11.2%) underwent reintervention, including reoperation (eight), balloon dilation (one) and intraoperative stents placement (one), for anastomotic (four) or diffuse PV stenosis (six), with four late deaths. Kaplan–Meier survival is 87.3 ± 0.036 SE% at 18.07 years with no difference according to anatomic type or surgical technique. Freedom from PV reintervention for operative survivors is 86.7 ± 0.052 SE% at 18.07 years. Cox proportional hazard indicates associated anomalies ($P = 0.008$) and reoperation for intrinsic PV stenosis ($P = 0.034$) as independent predictors of mortality. According to logistic analysis, preoperative obstruction predicts higher risk of reintervention for intrinsic PV stenosis ($P = 0.022$), while the double-patch technique increased the risk of late arrhythmias ($P = 0.005$). **Conclusions**: Side-to-side anastomosis provides excellent results for TAPVC repair while left atrial enlargement procedures appear to be associated with higher risk of late arrhythmias. Although early and aggressive reintervention for recurrent PV obstruction is mandatory, intrinsic PV stenosis remains a predictor of adverse outcome.

Keywords: Total anomalous pulmonary venous connection; Pulmonary vein stenosis

1. Introduction

Total anomalous pulmonary venous connection (TAPVC) is a rare, though heterogeneous anomaly, accounting for only 2% of congenital heart disease [1]. In some variants, it represents the only true surgical emergency in congenital heart surgery. The current remarkable results of TAPVC repair [2–4] reflect the general improvements in preoperative, intraoperative and postoperative management strategies which have occurred over the last decade. The aim of this study was a long-term retrospective evaluation of non-isomeric TAPVC repair at a single institution, identifying the risk factors associated with increased mortality and morbidity, while controlling for anatomic TAPVC subtype and technique of repair.

2. Materials and methods

2.1. Demographics and preoperative evaluation

Between August 1983 and August 2001, 89 consecutive patients (median weight 3.8 kg) underwent repair of non-isomeric TAPVC at the Bambino Gesù Children's Hospital in Rome. Median age at first operation was 54 days (range 2 days to 24.3 years). Median age at surgery significantly decreased to 24.5 days after 1992.

Echocardiography became the gold-standard diagnostic tool in 1988 and indication for surgery has been exclusively based on detailed echocardiographic study in the last 39/64 patients (60.9%). Cardiac catheterization was performed in

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Table 1
Anatomic diagnosis of TAPVC, related classification in obstructed versus non-obstructed lesions and overall mortality

Anatomic type	Number (%)	Obstructed (%)	Early mortality (%)	Late mortality (%)
Supracardiac	38 (42.7)	10 (26.3)	3 (3.4)	1 (1.1)
Cardiac	26 (29.2)	7 (26.9)	1 (1.1)	1 (1.1)
Infracardiac	16 (18.0)	14 (87.5)	2 (2.2)	2 (2.3)
Mixed	9 (10.1)	1 (11.1)	1 (1.1)	0 (0)
Total	89 (100)	32 (35.9)	7 (7.8)	4 (4.5)

the remaining 25 patients, mainly to confirm unusual or unclear patterns of pulmonary venous (PV) connection in mixed (six patients) or infracardiac TAPVC (six patients).

Anatomical diagnosis and related classification in obstructed versus non-obstructed lesions are depicted in Table 1. Obstruction was diagnosed when, in addition to poor clinical conditions, there was evidence of systemic or suprasystemic pulmonary artery pressures, associated with angiographic evidence of obstruction, or when echocardiography demonstrated acceleration in the common pulmonary vein. Associated anomalies other than PDA were diagnosed in ten patients (11.2%), including multiple VSDs (two), supramitral ring (one), Goldenar syndrome with right lung hypoplasia and subaortic VSD (one), complete (one) or partial (one) atrioventricular septal defect, the latter associated with muscular VSD and aortic coarctation, GI tract malformations with anorectal (one) or esophageal (one) atresia, diaphragmatic relaxation (one) and aortic coarctation (one). TAPVC repair was performed as an emergency procedure in 28 patients (31.5%), on an urgent basis (next available operating list) in six patients (5.6%) and electively in 55 patients (61.7%).

2.2. Operative technique

Repair was accomplished under deep hypothermia and circulatory arrest in 62 patients (69.6%). Continuous cardiopulmonary by-pass with cardioplegic cardiac arrest has been recently preferred in 27 patients (30.4%) with supracardiac (ten patients), cardiac (14 patients) and mixed (three patients) TAPVC. Median by-pass perfusion times were 105 min, cross-clamp 44 min and circulatory arrest time 42 min. Repair was accomplished by the double-patch technique with left atrium enlargement in 29 patients, side-to-side anastomosis in 29 patients, coronary sinus unroofing in 24 patients and a combination of the above in six patients. The double-patch technique [5] was most frequently selected for supracardiac (22 patients) and infracardiac (three patients) TAPVC repair until 1990. This technique (Fig. 1a) involves a transversal incision of the right atrium, with division of the crista terminalis, finally entering the dome of the left atrium. The PV confluence is longitudinally opened. The right side of the anastomosis between the PV sinus and the left atrium is patch augmented. Simultaneous left atrial enlargement is achieved by rightward shifting of the second patch used for inter-atrial septation. The first patch is then flipped over to complete right atrial enlargement. Since 1990, a superior approach in the inter-aortic caval space (Fig. 1b) as describer by Tucker et al. [6] and a 'tilted-up apex' technique as described by Phillips et al. [7] have been respectively preferred for supracardiac (16 patients) and infracardiac (13 patients) TAPVC repair. We defined the combination of the superior approach and the 'tilted-up apex' technique as a side-to-side anastomosis between the PV confluence and the left atrium. TAPVC of the cardiac type was repaired by coronary sinus unroofing with patch closure of the resulting defect. The incision on the roof of the coronary sinus was extended up to the junction with the PV confluence, in order to address any potential stenosis occurring at this level [8]. A combination of the above mentioned techniques was used to repair the mixed type of TAPVC. Delayed closure of the sternum (mean 2.6 days) was selected in 31 patients to accommodate normalization of right ventricular work and stabilization of pulmonary artery pressures.

2.3. Postoperative care and follow-up

Continuous monitoring of pulmonary artery or right ventricular pressure has been used since 1988, representing a useful parameter in the prevention of pulmonary hypertensive events, along with continuous paralysis and sedation, hyperventilation, use of vasodilators, bronchodilators and nitric oxide inhalation when indicated. Routine postoperative 2D-echocardiography and Doppler allowed visualization and flow patterns of the PV return, estimating right ventricular pressure and pressure gradients across the anastomosis between the PV confluence and the left atrium. Postoperative PV stenosis was diagnosed on the basis of PV flow patterns which did not reach baseline throughout the cardiac cycle, or documentation of estimated right ventricular pressures over 40 mmHg. Further investigation included trans-esophageal echocardiography, focusing on the morphology of the PV anastomosis, and cardiac catheterization, to visualize the course of the PVs and the extent of obstruction. After hospital discharge, all patients were followed at regular intervals (4 weeks, 3 months, 6 months, then yearly) at the outpatient clinic or by the local cardiologist. EKG, 24 h Holter and echocardiographic evaluations were available for review. After the third year of age, 28 patients underwent evaluation of their functional capacity by pulmonary function tests and exercise stress tests. Follow-up information was available for all but one hospital

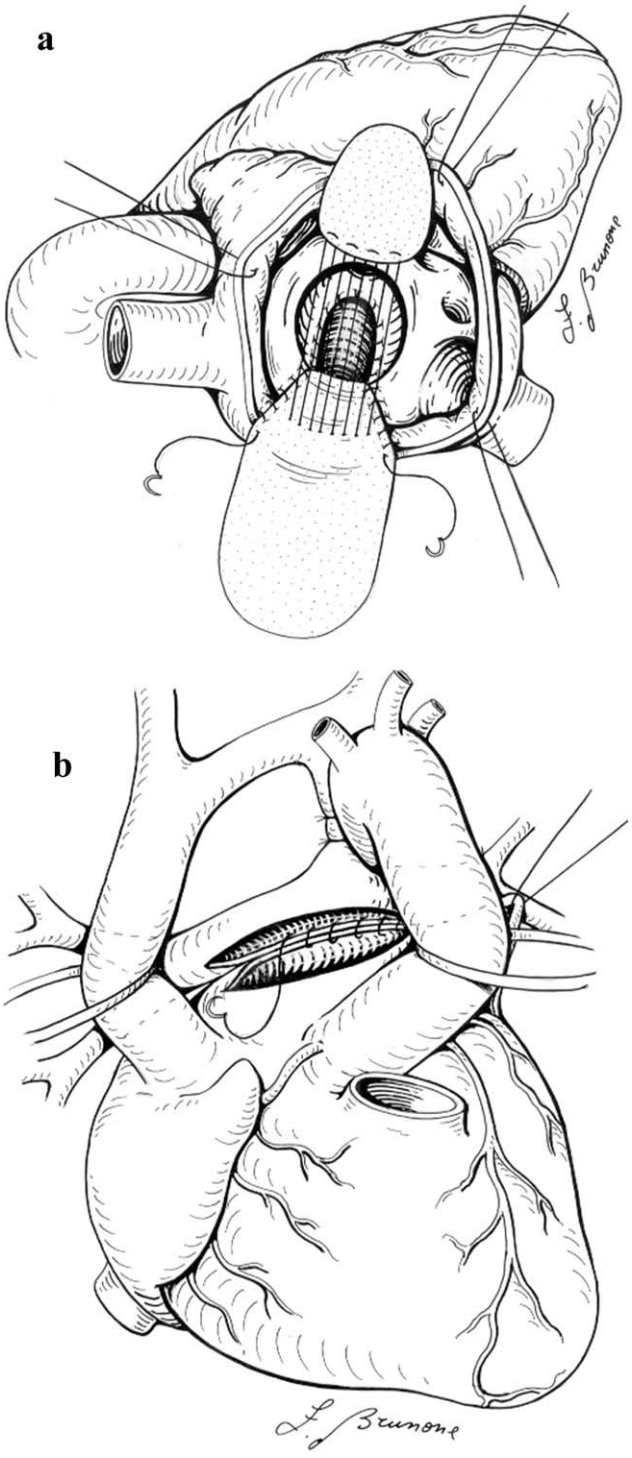

Fig. 1. (a) Double-patch technique. A transverse right atriotomy was performed. The incision is carried through the crista terminalis and across the foramen ovale, toward the back wall of the left atrium. The left atrial appendage is retracted. The anastomosis between the PV confluence and the left atrium is augmented with an heterologous pericardial patch. Left atrial enlargement is accomplished by rightward shifting of the second patch for inter-atrial septation. The first patch is finally flipped over to close the right atriotomy. (b) Superior approach for supracardiac TAPVC. The PV confluence and the dome of the left atrium are exposed through the inter aortic-caval space and a side-to-side anastomosis is accomplished without interposition of patching material.

survivor (77/78 patients, 98.8%). Total follow-up was 727.6 patient-years. Follow-up ranged from 0.7 months to 18.07 years (mean 8.86 years).

2.4. Statistical methods

Kaplan–Meier survival and freedom from reoperation were calculated using the SAS Statview-1998 statistical software (SAS Institute Inc., Cary, NC). Selected end-points were death, reoperation or interventional cardiologic procedure (balloon dilation or stent placement). Difference in survival was estimated by Log-rank testing. Dichotomous variables were analyzed by the χ^2 and Wilcoxon rank-sum tests. Variables associated with increased risk of early (30 day) death, late death and risk of reoperation for recurrent PV stenosis were assessed by univariate and multivariate logistic regression and by Cox proportional risk multivariate analysis.

3. Results

3.1. Mortality

Overall mortality was 12.3% (11/89 patients). Early (30 day) mortality was 7.86% (7/89) while late mortality was 4.5% (4/89). Causes of early death were: low output syndrome (two), combined low output and residual unilateral PV stenosis (one), pulmonary hypertensive event (one), cerebral hemorrhage (one), aortic hemorrhage (one) and Gram negative sepsis (one).

Low output syndrome was the main (two) or concurrent (one) cause of early death in three patients. The first one presented with an obstructed supracardiac TAPVC on the third day of life. The unusual echocardiographic diagnosis was further investigated by cardiac catheterization, which confirmed the connection of all PV to the superior vena cava at the junction with the azygous vein via an obstructed common trunk. The critical hemodynamic conditions forced an emergency TAPVC repair, which was accomplished by a side-to-side anastomosis. In spite of apparently good PV decompression, the patient could not be weaned from cardiopulmonary by-pass because of elevated left atrial pressure. Autopsy showed wide open anastomosis between the PV confluence and the left atrium, a diminutive LV with a mitral valve annulus of 8 mm in diameter, and splanchnic and pulmonary congestion with parenchymal pulmonary hemorrhage. The anatomic diagnosis in the second patient was obstructed TAPVC to the right atrium. The PV confluence was connected to the right atrium through an obstructive common trunk, which drained independently from the coronary sinus orifice. The patient had previously undergone an atrial septectomy on the 19th day of life. PV obstruction recurred and TAPVC repair was accomplished on the 29th day of life by side-to-side anastomosis of the PV confluence with the left atrium and patch augmentation of the right atrium. The patient could not be weaned from

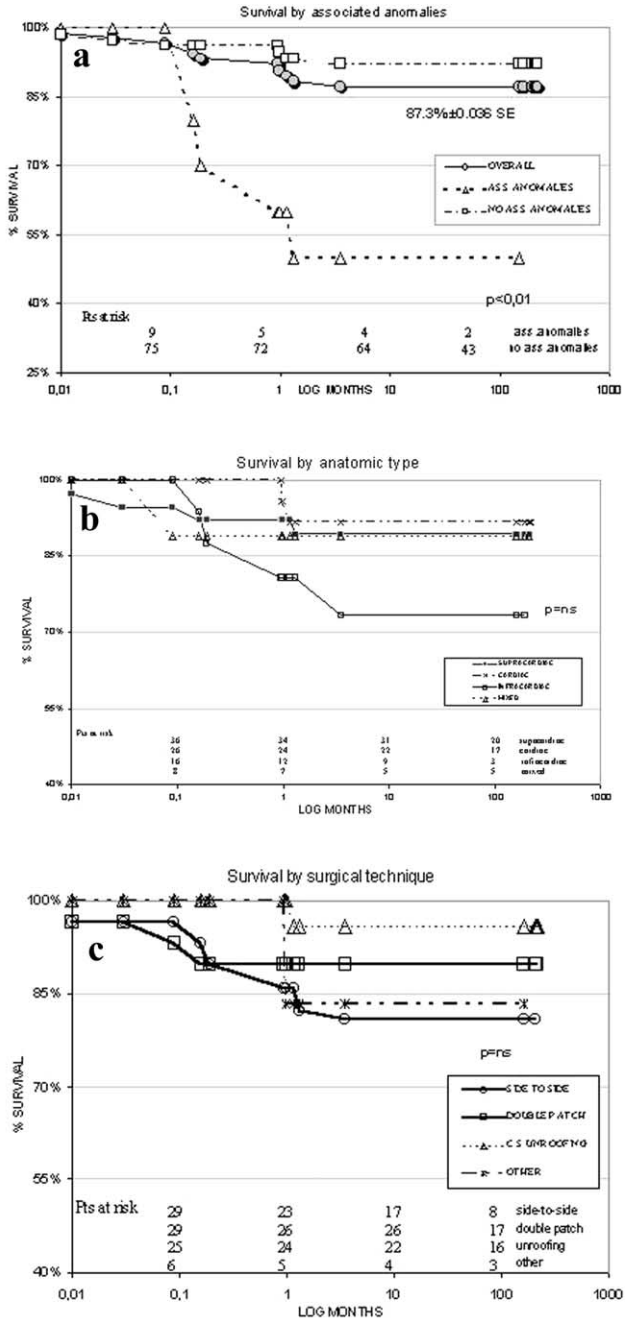

Fig. 2. (a) Actuarial survival at 18.07 years by the presence or absence of associated anomalies. (b) Actuarial survival by anatomic type of TAPVC. (c) Actuarial survival by type of surgical technique.

cardiopulmonary by-pass. Autopsy showed good reconstruction and moderate hypoplasia of the left-sided sections, with right ventricular hypertrophy. The anatomic diagnosis in the third patient was obstructed infracardiac TAPVC, partial atrioventricular septal defect, muscular VSDs and aortic coarctation. She underwent TAPVC repair by side-to-side anastomosis, patch aortoplasty and pulmonary artery banding on the second day of life. On the third postoperative day, persistence of low output and echocardiographic evidence of residual stenosis of the right pulmonary veins prompted reintervention. Right pulmonary vein plasty and PA band tightening were performed; nevertheless, the patient did not recover and eventually died the following day. Autopsy demonstrated adequate aortic arch reconstruction and good TAPVC repair in the presence of a diminutive left ventricle with mitral valve hypoplasia (5 mm diameter).

Pulmonary hypertensive crisis, cerebral hemorrhage and aortic hemorrhage were the causes of early death in three additional patients with infracardiac, mixed and supracardiac TAPVC. Autopsy was performed in all three. Histology of the pulmonary specimens in the first patient demonstrated the presence of increased medial thickness of both small pulmonary arteries and veins. One additional patient with supracardiac TAPVC and previous colostomy for anorectal atresia died on the fourth postoperative day for Gram negative sepsis, after an otherwise uncomplicated TAPVC repair by side-to-side anastomosis.

Causes of late death were right ventricular failure (one) and recurrent PV stenosis (three). All four deaths occurred after reoperation and are therefore discussed under the following section.

Overall actuarial survival was 87.3 ± 0.036 SE% at 18.07 years. Actuarial survival in patients with no associated cardiac or extracardiac anomalies other than TAPVC was 92.1 ± 0.031 SE% at 18.1 years (Fig. 2a).

TAPVC anatomy and type of surgical technique did not affect survival (Fig. 2b,c). Cox proportional hazard indicates the presence of associated anomalies to TAPVC and need of reoperation for intrinsic PV obstruction as independent predictors of mortality (Table 2). The presence of preoperative obstruction reached borderline significance ($P = 0.066$). A secondary analysis was conducted on the most prevalent group of patients, those with supracardiac TAPVC (38 patients), comparing results of the double-patch technique (22 patients) with those achieved by the superior approach (16 patients). Actuarial survival of supracardiac TAPVC repair by the two-patch technique was 89.7 ± 0.044 SE% at 18.1 years (mean 153 months), which is not significantly different from an 85.1 ± 0.097 SE% survival at 10.8 years (mean 52 months) achieved by a superior inter-aortic-caval approach ($P = 0.29$).

Table 2
Independent predictors of mortality according to the Cox proportional hazard method

Covariate	Estimate	SE	*P* value	Exp (coeff)
Associated anomalies	1.7	0.64	0.008	5.53
Intrinsic PV re-stenosis	1.69	0.66	0.010	5.46
Preoperative obstruction	1.2	0.65	0.066	—

3.2. Reoperation

Sixteen patients underwent an additional surgical procedure for recurrent PV stenosis (ten), VSD closure and PA

plasty (one), right hemidiaphragm plication (one) and pacemaker implantation (four).

3.2.1. Recurrent PV obstruction

Ten patients (11.2%) underwent reintervention because of recurrent PV stenosis. This was defined as extrinsic stenosis, when occurring at the level of the PV anastomosis (four patients), and intrinsic stenosis, when secondary to an endocardial thickening of the PV ostia (six patients) frequently resulting in diffuse sclerosis of the body of the PVs (three patients). A total of 12 procedures (1.2 per patient) were performed, including reoperation (nine), balloon dilatation (two) and intraoperative stent placement (one). The mean time interval between initial surgery for TAPVC repair and reoperation or cardiologic reintervention was 2.2 months in the case of intrinsic obstruction, and 9.7 months in the case of extrinsic obstruction ($P = 0.0169$). Extrinsic stenosis was cured (100% survival with no residual stenosis) by patch enlargement (three) or balloon dilation (one) of the anastomotic stricture. A variety of surgical techniques were used in the case of intrinsic PV obstruction, including patch augmentation (four), in situ pericardium (one) and longitudinal incision with transversal plication (two). Intraoperative placement of multiple stents in the PV ostia was attempted in one patient after unsuccessful patch augmentation of the PV anastomosis. Reintervention for intrinsic obstruction carried a 66% mortality rate (4/6), irrespective of the surgical technique. Autopsy showed increased medial thickness of both small pulmonary arteries and veins with endocardial thickening in three patients. Freedom from reoperation for intrinsic PV re-stenosis was 74.5 ± 0.1 SE% in the case of preoperative obstruction, compared with 98.2 ± 0.018 SE% in the case of no preoperative obstruction ($P < 0.001$) (Fig. 3a). Overall freedom from reoperation for any type of PV re-stenosis was 86.7 ± 0.052 SE% at 18.1 years (Fig. 3b). Logistic regression demonstrated no correlation between anatomy or technique of TAPVC repair and reoperation for PV stenosis, but confirmed a higher risk of recurrent intrinsic PV stenosis in the case of preoperative obstruction (Table 3).

3.2.2. Other reoperations

One patient with Goldenar syndrome, infracardiac TAPVC, right lung hypoplasia, conoventricular VSD and ostium secundum type ASD was reoperated 5 weeks after initial TAPVC repair by side-to-side anastomosis and banding of the pulmonary artery performed on the third day of life. Indication for reoperation was refractory mechanical ventilation dependency with evidence of left to right shunting (QP/QS 1.7:1), mean PA pressure of 12 mmHg and kinking of the left pulmonary artery take-off at cardiac catheterization. She underwent patch closure of the VSD, debanding and patch plasty of the pulmonary artery and closure of the ostium secundum type ASD with a fenestrated patch. She unfortunately died the following day due to right ventricular failure. Autopsy confirmed the diagnosis and demonstrated near-atresia of the right pulmonary veins.

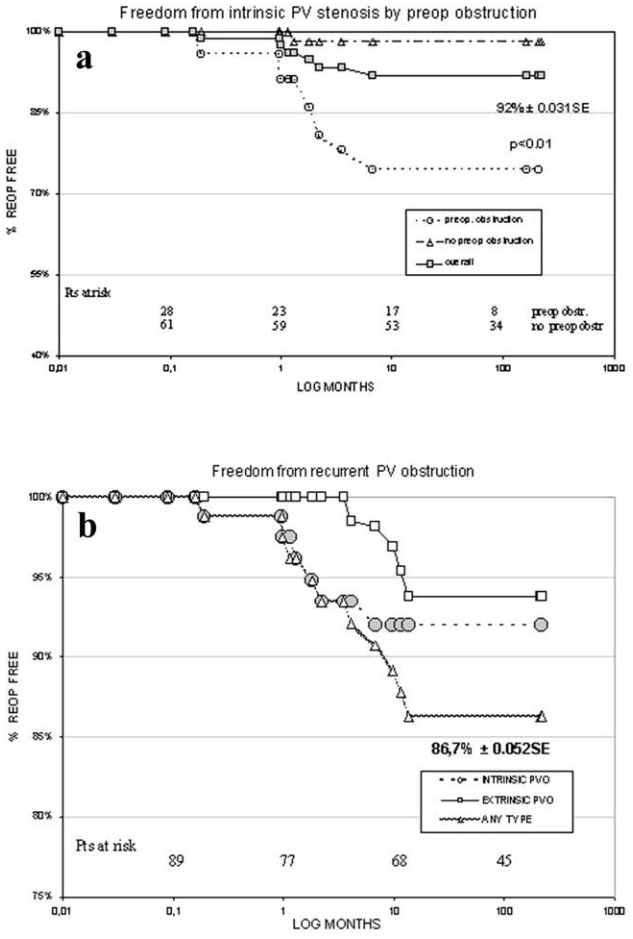

Fig. 3. (a) Actuarial freedom from reoperation for intrinsic PV re-stenosis by the presence or absence of preoperative obstruction. (b) Actuarial freedom from reoperation for PV re-stenosis by type or recurrent obstruction. Intrinsic obstruction occurred earlier and carried a worse prognosis than extrinsic obstruction.

One patient underwent right hemidiaphragm plication 11 days after repair of infracardiac TAPVC, with uneventful further clinical course. Four patients underwent pacemaker implantation at a mean time interval of 51.4 months from initial repair for supracardiac (two), cardiac (one) and infracardiac (one) TAPVC. There was no correlation between

Table 3
Intrinsic PV obstruction and late arrhythmias after TAPVC repair: risk assessment by the logistic regression method

Covariate	Estimate	SE	*P* value	Exp (coeff)
Reoperation for intrinsic pulmonary vein re-stenosis				
Preoperative obstruction	2.56	1.12	0.022	13.04
Late arrhythmias requiring medical therapy or pacemaker				
Double-patch technique	1.80	0.65	0.0050	6.3

the type of surgical technique and the need for pacemaker implantation. Indications for pacemaker implantation were symptomatic junctional bradycardia (two), AV block secondary to medical therapy for refractory atrial flutter (one) and third degree AV block (one).

3.3. Late outcome

Updated follow-up information was available on 77/78 survivors (98.8%). Currently, 72 patients are in functional class I on no oral medications (93.5%), while five patients are in functional class II. Beyond the ten patients who required reintervention, cardiac catheterization was repeated in three additional cases at a mean interval of 1.3 months from TAPVC repair to rule out recurrent PV stenosis. Pulmonary artery pressures were found to be half-systemic in a patient with multiple VSDs and residual left-to-right shunting at the ventricular level with QP/QS 1.3:1. Adequate TAPVC repair in the presence of half-systemic pulmonary artery pressures was confirmed by cardiac catheterization in a second patient 5 weeks following supracardiac TAPVC repair by the double-patch technique. Follow-up echocardiography documented a progressive fall in pulmonary artery pressures with resolution of the roentgenographic image suggestive of residual PV congestion. This scenario was interpreted as slow resolution of residual pulmonary lymphatic congestion. Normal pulmonary artery pressures were documented in the third patient with supracardiac TAPVC and stenosis of the right main bronchus. All the remaining 74 late survivors have no echocardiographic evidence of residual pulmonary hypertension.

Routine EKG and 24 h Holter follow-up evaluation demonstrated that the double-patch technique was associated with a 17.2% incidence of late arrhythmias requiring medical therapy or pacemaker implantation, including junctional bradycardia (three) or ectopic atrial rhythm (two). The mean time interval between two-patch TAPVC repair and onset of atrial arrhythmias was 54 months. The incidence of atrial arrhythmias was 3.4% in the case of side-to-side anastomosis, and 8.3% in the case of coronary sinus unroofing. No bradyarrhythmia occurred in the case of side-to-side anastomosis. Logistic regression demonstrated a higher risk of late arrhythmias in the case of repair by the double-patch technique ($P = 0.0050$) (Table 3). The functional capacity after TAPVC repair was evaluated after the third year of age in 28 late survivors. Pulmonary function tests and exercise stress tests showed normal vital capacity in 27 cases (96.4%) and normal exercise tolerance in 15 (55.5%).

4. Discussion

TAPVC is a relatively rare anomaly accounting for only 2% of congenital heart disease [1]. The anatomic patterns of anomalous PV connection are heterogeneous, since persistent splanchnic venous connections can occur at almost any point in the central cardinal or umbilicovitelline venous systems. The anatomical spectrum of TAPVC therefore reflects a wide difference of physiology and clinical presentation in postnatal life, ranging from right-to-left shunting with mild cyanosis to pulmonary edema. Obstruction of the PV pathway is a powerful predictor of adverse natural outcome and the tendency for PV obstruction in the infracardiac type of TAPVC is well recognized [9], especially when the pattern of infracardiac connection prevents the ductus venosus from by-passing the liver [10]. Nevertheless, the natural history of TAPVC is unfavorable per se, with a 50% mortality in the first 3 months of life and a median survival of approximately 2 months [11]. Therefore, early surgical repair is currently recommended, even before the onset of clinical symptoms and irrespective of anatomic subtype [2,3]. Results of TAPVC repair in infancy have markedly improved in recent years, with an operative mortality of 5% or less in some institutions [2–4,6]. This improvement is probably multi-factorial, mainly due to early non-invasive diagnosis and aggressive preoperative, intraoperative and postoperative management. Routine use of echocardiography as the gold-standard diagnostic tool, improvements in myocardial protection with specific attention to protection of the right ventricle, creation of a large and tension-free anastomosis with maximal use of the venous confluence and atrial tissue, careful geometric alignment of the PV sinus with the body of the left atrium avoiding torsion and rotation of the pulmonary veins, monitoring and prevention of pulmonary hypertensive events and delayed sternal closure have probably played a major role in reducing operative mortality. Risk factors like venous obstruction on presentation, urgency of operative repair and infradiaphragmatic anatomical type are no longer correlated with early mortality [12]. Our 92.1 ± 0.031 SE% 18.1 year survival for TAPVC repair in the absence of associated cardiac or extracardiac anomalies favorably compares with the experience of others [13], although we did not see a dramatic change in early mortality in recent years. This may be partly explained by the non-uniform surgical strategy adopted over the past 18 years. At the beginning of our experience, we were intrigued by the experimental work of Goor [14] on the close correlation between small atrial volumes and a decrease in cardiac output in TAPVC. Increased atrial compliance was thought to improve cardiac performance, and a two-patch technique with left atrial enlargement was routinely selected until 1990 for supracardiac and infracardiac TAPVC repair. Since 1990 we have preferentially adopted a superior approach through the inter-aortic caval space for supracardiac TAPVC [5] and since 1993 a 'tilted-up apex' technique for infracardiac TAPVC [6], therefore avoiding any patching material. This change in surgical strategy was prompted by the observation that adequate left atrial enlargement and volumes may result from the incorporation of the common PV sinus into the left atrium [14]. Results with the two-patch technique

have been satisfactory, but not significantly different from those obtained with the superior approach for supracardiac TAPVC. Moreover, logistic regression demonstrated that the double-patch technique was associated with a higher risk of atrial arrhythmias, including both bradyarrhythmias and tachyarrhythmias. Since the mean time interval between TAPVC repair by the two-patch technique and onset of atrial arrhythmias was comparable to the mean follow-up of the patients undergoing TAPVC repair by side-to-side anastomosis, it seems reasonable to assume that onset of atrial arrhythmias is indeed correlated with the technique of repair. The double-patch technique involves a transversal incision of the right atrium, with division of the crista terminalis, finally entering the dome of the left atrium. Division of the crista terminalis and surgical reconstruction by extensive use of patching material, with potential re-entry phenomena, may be responsible for the higher risk of atrial arrhythmias seen with this type of technique.

Despite the excellent results in early survival in most reported series, the incidence of reoperation for progressive PV obstruction is not negligible, ranging from 9 to 11% irrespective of the surgical technique [13,15,16]. Mortality for this complication varies between 30 and 45% [13,16], and alternative catheter interventions do not offer definitive solutions. Hyde [13] reported an 11% reoperation rate for PV stenosis on a cohort of 85 patients undergoing TAPVC repair over a 10 year interval. Despite initial improvements with non-surgical interventions (mean 2.4 per patient), PV obstruction reoccurred within 4 weeks in all patients, requiring 23 further surgical procedures (mean 2.5 per patient). Mortality at reoperation was 44%, with only 2/5 survivors completely free from residual stenosis. Lacour-Gayet [16] reported a 9% reoperative rate for recurrent PV obstruction in a series of 178 patients over a 15 year interval. Obstruction occurred 4 months after initial TAPVC repair. Poor results were experienced with non-surgical interventions, and the 16 patients underwent a total number of 20 reoperations (mean 1.25 per patient) with 31% mortality. Sutureless in situ pericardium repair was effective in relieving stenosis, especially when the right-sided PVs were involved.

In our experience, reoperation for PV obstruction was a powerful predictor of adverse outcome. The impact of preoperative PV obstruction as an independent risk factor of early mortality has been neutralized by changes in preoperative, operative and postoperative care [11], nevertheless this substrate might still play a role in the onset of recurrent PV obstruction. Recurrent PV obstruction can be localized at the site of the PV anastomosis (extrinsic obstruction) or it may be secondary to endocardial thickening of the PV ostia frequently resulting in diffuse PV sclerosis (intrinsic obstruction). Intrinsic obstruction occurs earlier and carries a worse prognosis [13,16]. Our experience validates this observation. The mean time interval between TAPVC repair and reoperation or cardiologic reintervention was indeed 2.2 months in the case of intrinsic obstruction and 9.7 months in the case of extrinsic obstruction ($P = 0.0169$). Extrinsic stenosis was always cured by patch enlargement or balloon dilation of the anastomotic stricture, while intrinsic obstruction carried a 66% mortality rate, irrespective of the surgical technique (patch augmentation, in situ pericardium or longitudinal incision with transversal vein plasty). Preoperative obstruction predicted a higher risk of intrinsic PV re-stenosis in our cohort. As a logical deduction, the type of suture material used for the anastomosis between the PV confluence and the left atrium did not appear to have an impact on the occurrence of intrinsic obstruction. Previous anatomic studies [17] have shown that preoperative PV obstruction is associated with increased medial thickness in both pulmonary arteries and veins, which exceeds by far the degree of medial changes observed in other lesions with large QP/QS and pulmonary hypertension. Based on our limited experience, we speculate that the medial and intimal changes seen in preoperative obstruction may predispose to the development of intrinsic PV stenosis, even in the presence of adequate PV decompression.

In conclusion, excellent long-term survival and reoperation freedom are expected with early repair of TAPVC, especially in the absence of associated cardiac or non-cardiac anomalies. We currently favor a tissue-to-tissue anastomosis (side-to-side technique) to reduce the risk of late arrhythmias seen with extensive use of patching material. The presence of preoperative obstruction appears to predict a higher risk of intrinsic PV re-stenosis. Although early and aggressive reintervention for recurrent PV stenosis is mandatory, intrinsic obstruction remains a predictor of adverse outcome.

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