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CLINICAL STUDY

Long-term results of surgical treatment of total anomalous pulmonary venous drainage in children

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Abstract: *Aim of study:* Retrospective analysis of surgical correction of TAPVD performed between January 1992 and March 2008.
Methods: Review of patients' medical records. Patients' preoperative, operative as well as postoperative data were collated and analyzed using JMP statistical program version 5.
Results: A total of 51 patients with total anomalous pulmonary venous drainage underwent surgery at our center during a period of over seventeen years. Actuarial survival was 90.2 %. Early postoperative death was recorded in 4 patients (7.8 %) as against one late postoperative death. The only statistically significant risk factor for death was the time of surgical repair. Patients undergoing the repair before 1997 were more likely to die than those operated on after this period, p=0.006. Patients' survival following the surgical correction prior to the year 1997 was 63.63 % as opposed to 97.5 % for the period between 1997 and 2008. Freedom from surgical re-intervention over the period of follow-up was 92 %. The obstructive type of TAPVD was associated with longer ICU stay and higher postoperative complications, p=0.003
Conclusion: We have recorded a significant improvement in patients' survival following surgery for total anomalous pulmonary venous drainage in the last decade. This can be attributed to a number of new measures both surgical and medical employed in the treatment of our patients *(Tab. 3, Fig. 7, Ref. 17).* Full Text (Free, PDF) *www.bmj.sk.*
Key words: TAPVD, total anomalous pulmonary venous drainage, obstruction of pulmonary vein.

Definition

Total anomalous pulmonary venous drainage is a cardiac malformation in which there is no direct connection between any pulmonary vein and the left atrium but, rather, all the pulmonary veins connect to the right atrium or one of its tributaries, anomalies in which the pulmonary veins drain into the systemic venous circulation via persistent splanchnic connection (1, 2). It is an infrequent cyanotic congenital heart disease accounting for about 1 % of all congenital heart diseases (3).

Classifications

The classification is based on prenatal errors in embryologic development.

There are 4 forms of TAPVD – supracardiac, cardiac, infracardiac and mixed type. Their names refer to the site where the pulmonary veins drain to the systemic venous circulation (4). The presence of obstruction within the pulmonary veins is an important hemodynamic factor. Hence, TAPVD has also been classified as obstructive (with pulmonary vein stenosis) and non-obstructive (without stenosis of pulmonary veins) (Fig. 1).

Surgical correction

The primary principles of surgical correction are to establish a non-obstructed communication between the common pulmonary vein and the left atrium, to interrupt the connections with the systemic venous circulation, and to close the atrial septal defect. The specific surgical technique is dependent on the type of anomalous connection and the presence of obstruction (5, 6, 7).

Aim of study

Our study is a retrospective analysis of surgical correction of TAPVD performed between January 1992 and March 2008 with the aim of:

1) Identifying the factors affecting the survival and good surgical outcome,

2) Identifying the risk factors for surgical re-do/re-intervention.

Methods

Patients' medical records were reviewed retrospectively. Preoperative as well as postoperative data were retrieved and statistically analyzed using the JMP statistical program version 5. The

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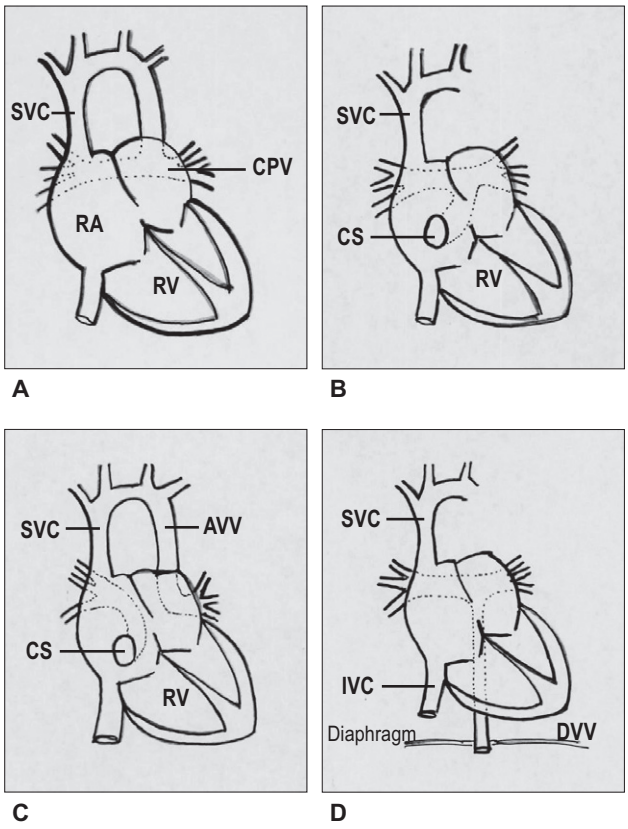

Fig. 1. Morphology and anatomy of TAPVD, A – supracardiac, B – cardiac, C – mixed, D – infracardiac types of TAPVD. Abbreviations: SVC = superior vena cava, IVC = inferior vena cava, RA = right atrium; RV = right ventricle, AVV = ascending vertical vein, DVV = descending vertical vein, CPV = common pulmonary vein.

Kaplan-Meier technique was used to determine overall patients' survival as well as freedom from re-intervention. Risk factors were determined using univariate analysis. The analysis of variance (ANOVA) technique was employed with continuous data while contingency analysis was used in the case of nominal data. A "P" value of less than 0.05 was accepted as statistically significant.

Results

A total of 51 patients underwent surgery for isolated form of TAPVD total 51. There were 17 (33.3 %) females and 34 (66.6 %) males. M:F = 2:1. 22 (43.13 %) patients had the supracardiac type of TAPVD, 6 (11.76 %) patients had the cardiac type, 16 (31.37 %) patients had the infracardiac type while 7 (13.72 %) patients had a mixed type of TAPVD (Tab. 1). 20 (39.2 %) patients had some degree of pulmonary vein obstruction. The highest rate of pulmonary vein obstruction was observed in patients with the infra-cardiac type of TAPVD. 75 % of all patients with the infra-cardiac type of TAPVD had pulmonary vein obstruction.

Tab. 1. Age distribution of all operated patients.

Age distribution (n)	51
Up to 1 month	30
1 to 3 months	10
3 to 12 months	9
Over 12 months	2

The mean age of patients at surgery was 62.62 days (±14 days). In patients with obstructive type of TAPVD the mean age at operation was 88.9 days (±17 days) while in patients with non obstructive type the average age was 21.85 (±21 days) (Fig. 2).

The mean ventilation time in all patients was 71 hours (±10 hours). Patients with the obstructive type of TAPVD had a mean ventilation time of 100 hours (±15 hours) as opposed to 53 hours (±12 hours) for those without obstruction. The average length of ICU stay was 6.2 days (±4.8 days). 7.7 days (±1.05 days) for the obstructive type and 5.2 days (±0.8 days) for the non-obstructive type (p=0.07). On the whole, a total of 8 (15.68 %) patients had delayed chest closure. 6 (75 %) of these patients were of the obstructive type.

Patients' survival

Survival: We recorded a 90.2 % actuarial survival over the period of follow-up. Of the total number of 51 patients, 5 deaths (9.8 %) were recorded. 4 (7.8 %) of the deaths occurred in the early postoperative period. Figure 3 shows the Kaplan-Meier survival curve for our patients' population.

Analysis of the likely causes of death revealed that:

2 patients died from severe pulmonary hypertension, 1 patient died due to dysfunction of the left ventricle, 1 died due to severe pulmonary vein obstruction and another died of bronchitis.

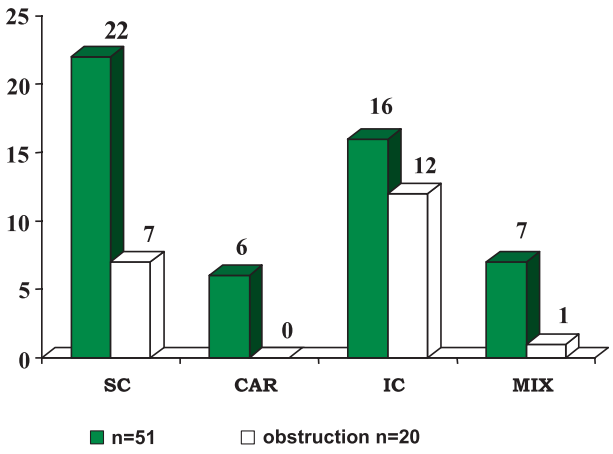

Fig. 2. Distribution of various types of TAPVD in our patients. SC – supracardiac type, CAR – cardiac type, IC – infracardiac type, MIX – mixed type of TAPVD.

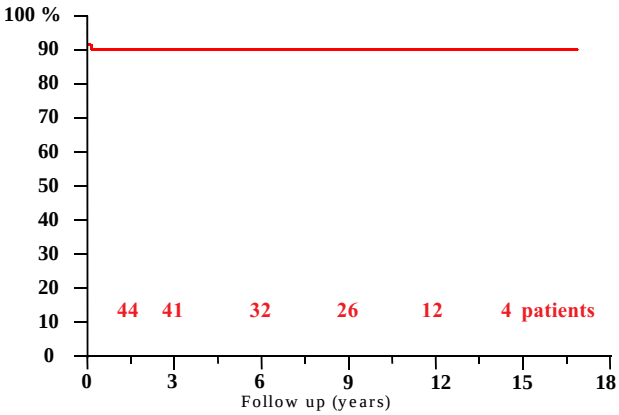

Fig. 3. Kaplan-Meier survival curve for our group of patients.

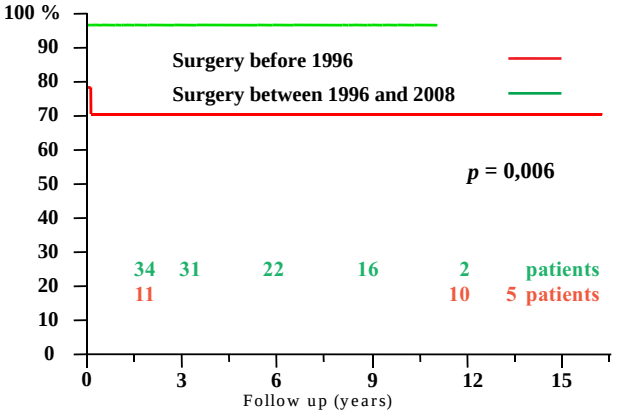

Fig. 4. Patient survival according to period of surgery.

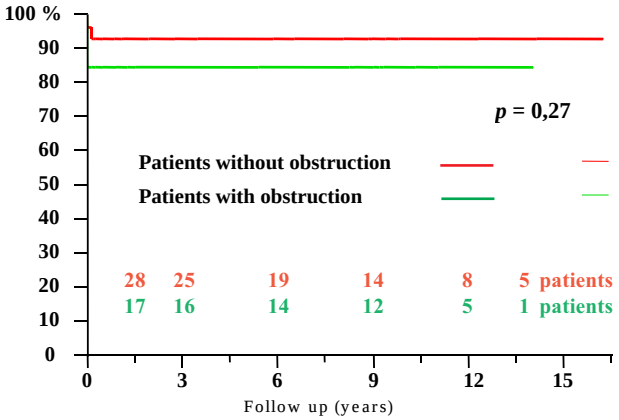

Fig. 5. Kaplan-Meier survival curve comparing the obstructive and non-obstructive types of TAPVD.

Risk factors for death: The only statistically significant risk factor for death was the time (year) of surgical repair. Patients undergoing repair before 1996 were more likely to die than those operated on after this period, p=0.006 (Fig. 4). Survival after surgical correction between 1992 and December 1995 was 63.63 %. In comparison, postoperative survival between January 1996 and March 2008 was 97.5 %. In other words, of the five deaths recorded over the period of follow-up, 4 occurred in those operated on before 1996.

The presence of pulmonary vein obstruction was not a significant risk factor for death in our series. Only 1 (20 %) of the 5 patients who died after surgery had some degree of pulmonary vein obstruction (Fig. 5).

Re-intervention: 4 (7.8 %) patients underwent surgical re-do due to obstruction of the pulmonary veins. This equates to a 92.2 % freedom from re-intervention over the period of follow-up. 3 of these patients undergoing surgical re-do had significant obstruction at the site of pulmonary vein anastomosis while 1 patient had a large thrombus in between the confluence of the pulmonary veins and the left atrium (Fig. 6).

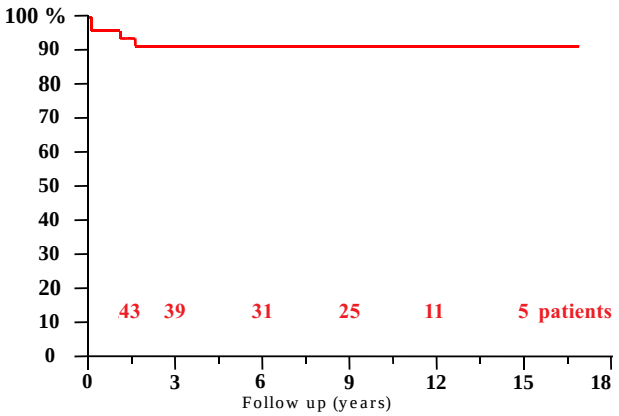

Fig. 6. Freedom from surgical re-intervention.

Risk factors for postoperative morbidity: Statistical analysis of factors affecting prolonged ICU stay and thus also postoperative morbidity showed a significant difference between the obstructive and non-obstructive types of TAPVD. Patients with the obstructive type of TAPVD had a higher risk of surgery-related complications than those without obstruction (p=0.003). As shown in Figure 7 below, 12 (60 %) of the 20 patients with obstruction had significant postoperative complications as against 6 (19 %) of 31 patients without obstruction.

Table 2 shows a list of some surgery-related variables as they occurred in the obstructive and non-obstructive subtypes of TAPVD.

Other postoperative complications: Table 3 shows a distribution of the most frequent postoperative complications as recorded in our series.

Limitations: Our study is limited by its retrospective nature and the relatively small size of the cohort.

Discussion

The outcome of surgery for isolated TAPVD has improved markedly over the years. In the early years of surgical repair of TAPVD, patients' survival was as low as 20 to 40 %. This figure improved to about 80 % in the late 70's and early 80's (8, 9).

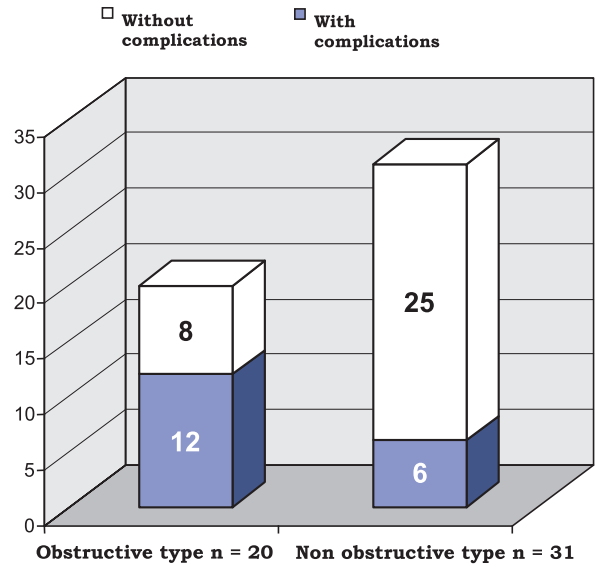

Fig. 7. The incidence of surgery-related complications in the obstructive and non-obstructive types of TAPVD.

In our series, patients' survival was over 90 % with early postoperative death at 7.84 %. In some renowned centers, mortality following surgical correction of isolated TAPVD is put at less than 15 % (5, 6, 7). In the non-obstructive subtype, postoperative mortality is about 5 %. We recorded a mortality rate of 6.45 % in the non-obstructive subtype. The picture looks a bit different in patients with pulmonary vein obstruction, where we recorded 15 % mortality. Higher rates of mortality have been reported in patients with TAPVD and other complex cardiac anomalies. However, such patients were not included in our present study.

The overall improvement in patients' survival as recorded in our series can be attributed to a number of new improved measures in healthcare. These measures which are at the level of preoperative, operative and postoperative management include:

Tab. 2. List of surgery-related variables as they occurred in the obstructive and non-obstructive.

Surgery-related variables	Obstructive type n=20	Non obstructive type n=31	p
mean ventilation time/hours	100	53	0.023
mean age/day	21	89	0.019
length of ICU stay/day	7.7	5.2	0.077
delayed chest closure	6	2	0.063
low cardiac output	9	3	0.003
postoperative obstruction of pulmonary veins	2	3	
pulmonary hypertension	9	6	0.049
Re-do	1	3	
postoperative death	3	2	
postoperative arrhytmia	16	20	0.33

Tab. 3. The most frequent complications following surgery for TAPVD.

Complications	n
Arrhytmia	37
Sepsis 11	
Multi organ failure	9
Open chest	9
Diaphragm plication	5
Chylothorax	2
Mediastinitis	2
Wound Re-suture	2

Quick and reliable diagnostic methods, improved ICU care ensuring that patients are stabilized prior to surgery, intra-operative measures aimed at better myocardial protection, better CNS protection and effective weaning from bypass (deep hypothermia, cold blood cardioplegia, modified ultra-filtration, etc), better postoperative ventilation and therapeutic options in the ICU.

Another area of interest to our study was recurrent pulmonary stenosis. Recent studies (2) have looked at the incidence of recurrent pulmonary stenosis vis-á-vis surgical techniques, types of suture material and the histology of the affected pulmonary veins. Reports from centers in Boston and London (2) put the incidence of recurrent stenosis following surgery for TAPVD at between 10 and 30 %. Only 4 (7.8 %) patients in our series required re-intervention due to recurrent pulmonary stenosis. As a group, pulmonary vein stenoses have a very poor prognosis. The congenital lesions can be classified as to whether the pulmonary veins are diffusely hypoplastic, long-segment focal (tubular) stenosis, or ostial (discrete) stenosis. The patients with diffuse hypoplasia of pulmonary veins that extends into the periphery of the lung have a very poor prognosis, and often the only therapy is heart-lung or lung transplantation (13). The patients with tubular long-segment hypoplasia of the pulmonary veins with reasonably normal intraparenchymal pulmonary veins also have a poor prognosis, but occasionally operative repair can result in long-term survival (10, 15, 16). In our series we did not record any cases of diffuse or tubular hypoplasia of pulmonary veins. Of the 4 patients undergoing surgical re-do due to obstruction of pulmonary veins 1 patient had the supracardiac, 1 had the cardiac, another had the infracardiac while the last patient had the mixed type of TAPVD. 3 of the cases were discrete (ostial) stenoses near the suture line while the fourth case was a large thrombus between the venous confluence and the left atrium. Recent studies have tried to analyze the impact of surgical techniques and suture materials on the incidence of recurrent pulmonary vein stenosis (11, 12). Some authors have advocated the use of the so-called "sutureless" technique even in primary repair of TAPVD (10, 11, 12) while according to others the use of absorbable sutures may reduce the formation of fibrous tissue around the suture line and thus prevent the postoperative obstruction of pulmonary veins (12). While little improvement has been made with the use of the "sutureless" technique, results

have yet to show whether there is any advantage of absorbable suture over non-absorbable suture (10). In our series the "sutureless" in-situ pericardial repair technique was employed in only one of the "re-do" cases while reconstruction of the pulmonary vein – left atrium anastomosis was carried out in the rest.

Another important technical question in repair of TAPVD is whether or not the vertical vein should be ligated. Many centers prefer to leave the vertical vein patent especially when there is an associated hypoplasia of the left heart structures or obstruction of the pulmonary vein (13, 14, 17). It has been argued that in the course of time, the vertical vein just like the patent ductus arteriosus will obliterate spontaneously. In our group of patients, the vertical vein was left patent only in 3 patients with supracardiac TAPVD, of whom 2 were of the obstructive type. Otherwise, we have routinely ligated the vertical veins. There have also been case reports of ischemia of the liver following the ligation of the descending vertical vein in patients with infracardiac TAPVD (13, 14). We did not record any cases of ischemia of the liver or other complications in connection with the ligation of vertical veins.

Our study has shown a higher incidence of postoperative pulmonary hypertension and increased lung reactivity particularly in patients with the obstructive type of TAPVD. These conditions have, in most cases, been successfully managed at ICU by adjusting the ventilation parameters and by the use of medications such as Sildenafil. Nitric oxide (NO) has been employed in such cases in our center since 1998 and Sildenafil since 2006.

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