The Turkish Journal of Pediatrics
Volume 27, No. 2, April - June 1985

TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE*

Surgical Correction

*Tayyar Sarıoğlu MD***, Şencan Özme MD***, Öztekin Oto MD****, Yurdakul Yurdakul MD*****, Mete Alp MD****, Sema Özer MD******, Aydın Aytaç MD******

Key words : anomalous pulmonary venous drainage, congenital heart disease

Anomalous pulmonary venous drainage (APVD) is a congenital cardiac malformation, in which pulmonary veins drain directly or indirectly into the right atrium, instead of the left atrium. Partial and total anomalous pulmonary venous drainage (TAPVD) are considered as two types of this malformation. TAPVD can be of the supracardiac type, where pulmonary veins drain into the right atrium via the superior vena cava or its tributaries; of the cardiac type (via coronary sinus or directly to the right atrium); or of the infracardiac type, where pulmonary venous return is through the inferior vena cava or portal vein[1,2].

TAPVD accounts for between 1% and 4% of all congenital cardiac defects[3,4]. Approximately 75% of patients with TAPVD die before the age of one year, the remainder usually die before reaching adulthood, if they are not treated surgically[5,6], but today, with modern cardiac surgical techniques total correction of TAPVD has become possible.

This report presents five TAPVD cases (cardiac and supracardiac types) on which, from the physiological point of view, total correction operations have been performed.

* From the Department of Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine, Ankara.

** Lecturer in Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine.

*** Professor of Pediatrics and Pediatric Cardiologist, Hacettepe University Institute of Child Health.

**** Resident in Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine.

***** Professor of Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine.

****** Pediatrician, Hacettepe University Faculty of Medicine.

Material and Methods

From August 1976 to November 1983, in the Department of Thoracic and Cardiovascular Surgery of Hacettepe University, total correction operations were performed on five TAPVD cases (four supracardiac, one cardiac). In one of the supracardiac cases, all of the pulmonary veins were returning to the right atrium via the innominate vein except for the left inferior pulmonary vein which was communicating directly into the left atrium. This patient has been included in the supracardiac type of TAPVD, because of the clinical, hemodynamic and surgical similarity to the other TAPVD cases.

Our patients were between five and seven years of age the mean age being six. Two of the patients sought medical advice on account of cyanosis and exercise intolerance while the others came with dyspnea on exertion and growth retardation. One of the cases was symptom free and was diagnosed on a routine medical examination.

Physical examination of the patients revealed fixed splitting of second heart sound and a systolic ejection murmur along the left sternal border. In one of our cases, the pulmonic component of the second heart sound was loud and accentuated.

The hematocrit level was 52% in a mild cyanotic case while the others had normal hematocrit levels. Electrocardiograms (ECG) revealed sinus rhythm, right axis deviation, right ventricular hypertrophy and in one case, right bundle branch block (RBBB) as well. Telecardiograms showed enlarged heart shadow and increased broncovascularity in all cases but one. In the supracardiac types, the "snowman" sign was also found in the chest X-ray (Figure 1). All the patients underwent cardiac catheterization and in four cases this revealed the supracardiac type of

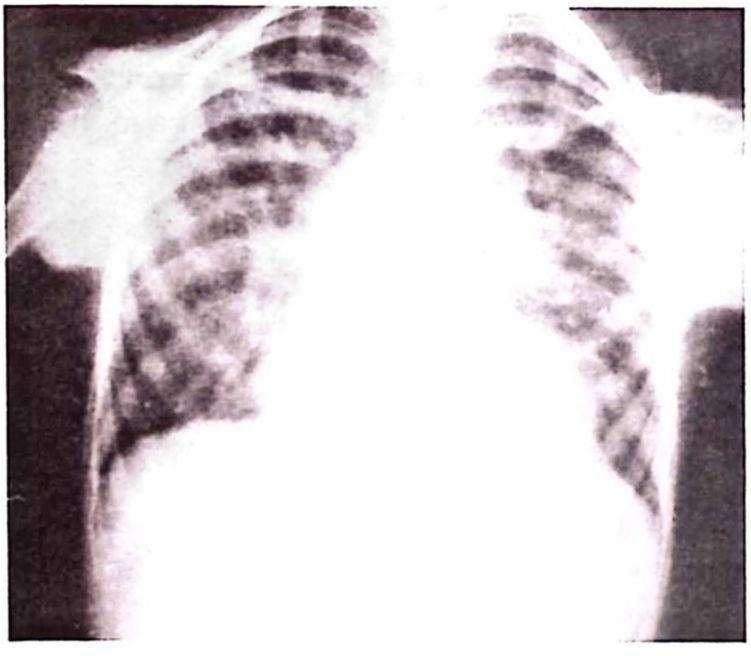

Figure 1

Snowman sign on chest X-ray

TABLE I

Clinical Data on Five Patients with TAPVD

Cases	Age	Sex	Type	Surgical technique*
B.C.	7	Male	Cardiac	Atrial septation
İ.U.	6	Female	Supracardiac	Right transatrial
M.D.	7	Male	Supracardiac	Posterior extracardiac
K.C.	5	Female	Supracardiac	Right transatrial
M.K.	7	Male	Supracardiac	Posterior extracardiac

*: See text

TAPVD (Table I). The pulmonary veins were first communicating with a common sac which was opening into the left innominate vein through a vertical vein (Figure 2). In addition, there was in all cases atrial septal defect (ASD). One of the cases underwent operation with the diagnosis of ASD and cardiac type TAPVD was detected peroperatively.

Median sternal split and a routine cardiopulmonary bypass technique were used on all patients. After the year 1978, 26 - 28 °C of hypothermia, cold K^+ cardioplegia and topical myocardial cooling were used while the cardiopulmonary

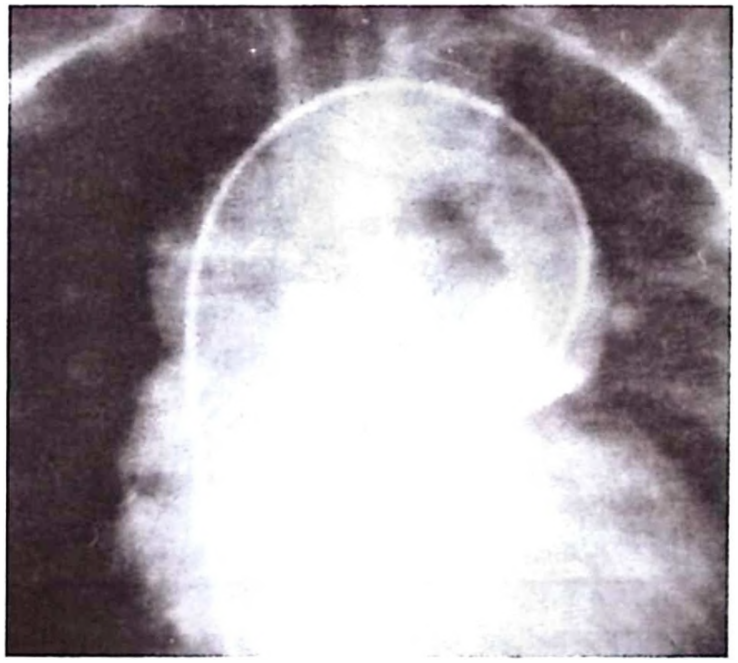

Figure 2

Angiographic appearance in supracardiac type of TAPVD. Note that the tip of the catheter is against the common pulmonary venous sac where the dye is given.

bypass was performed. In two of the supracardiac type cases, anastomosis between the common pulmonary venous sac and the left atrium was performed by an extracardiac approach while the apex of the heart was elevated. In these cases ASD was closed primarily. Also, the common vertical vein was ligated at the junction of the innominate vein.

In the other two supracardiac TAPVD cases, the transatrial approach was preferred[7]. After a total bypass, a transverse incision was carried out into the right atrial wall, and this was extended to the interatrial septum and into the left atrial wall and up to the left atrial appendix, posteriorly. The common pulmonary sac, which was located just behind the atria, was incised parallel to the previous atrial incision and the sac anastomosed to the posterior wall of the left atrium supplying a passage 4–6 cm in diameter (Figure 3). In one of these cases ASD was closed primarily, while in the second case the left atrium was enlarged by deviation of the interatrial septum with a pericardial patch (Figure 4). Consequently, the right atrium, which had become smaller, was also enlarged upon its anterior wall by another pericardial patch.

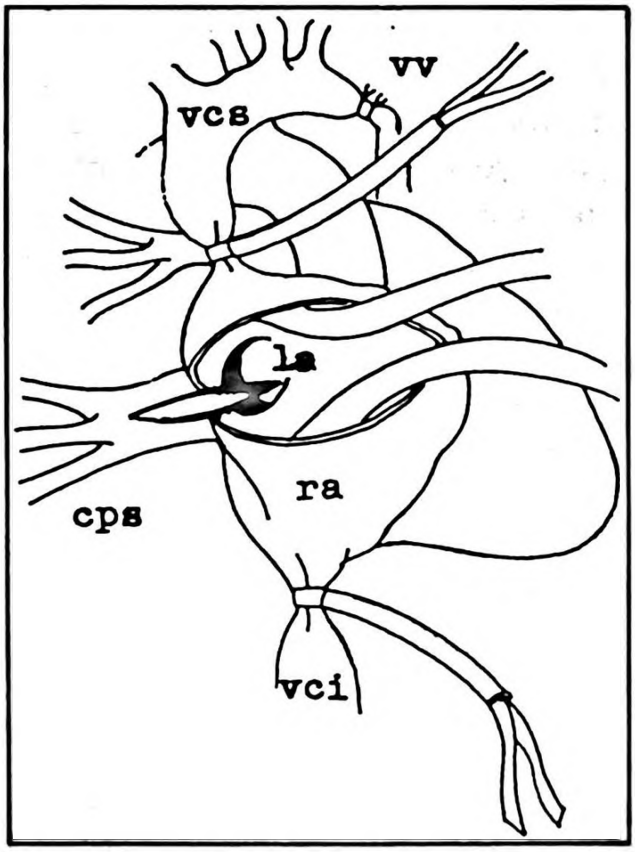

Figure 3

Repair of supracardiac TAPVD. Note the incision extending towards the posterior atrial wall.
cps = common pulmonary sac, vcs = vena cava superior, vci = vena cava inferior, ra = right atrium, la = left atrium, vv = vertical vein.

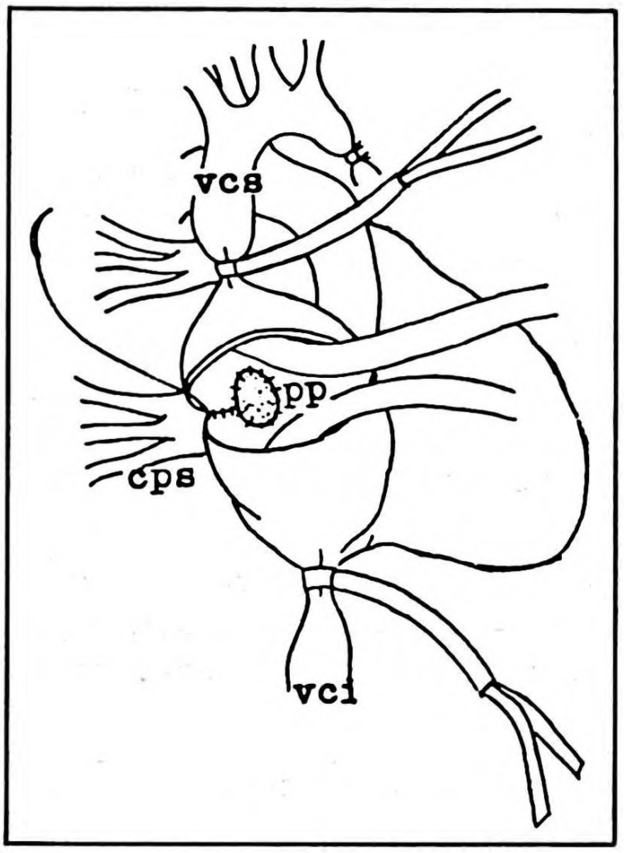

Figure 4

Closure of ASD with a pericardial patch.
cps = common pulmonary sac, vcs = vena cava superior,
pp = pericardial patch, vci = vena cava inferior.

In the cardiac type TAPVD case, there was a large ASD and all of the pulmonary veins were opening into the right atrium via a common trunk. The posterior wall of the left atrium was incised near the left upper pulmonary vein so that communication between this common trunk and the left atrium could be established. Then the ASD was closed with a large pericardial patch. Thus an effective drainage was supplied from the common pulmonary trunk to the left atrium.

Results

One patient only died from pulmonary venous congestion in the early postoperative period. In this case, the anastomosis between the left atrial appendix and the common pulmonary sac had been performed by an extracardiac approach. No complication was detected in the other cases, during either the early or the late postoperative period. The four patients who survived have been followed up for between one to eight years (mean 3.2 years). According to the New York Heart Association (NYHA) classification, they are in class I.

Discussion

TAPVD is a congenital malformation in which none of the pulmonary veins directly communicate with the left atrium[8]. Three types of TAPVD are recognized, known as the cardiac, the supracardiac and the infracardiac types, and among these the supracardiac type is the most frequent. Very occasionally two types of TAPVD are to be found in the same patient (mixed type)[2]. In the majority of supracardiac TAPVD cases, left and right pulmonary veins compose a common pulmonary sac behind the atrium. From this sac pulmonary venous drainage passes through another large vein called the vertical vein. This vein drains into the superior vena cava via the innominate vein. Usually, the mixing of the blood occurs at the atrial level by means of an ASD, and systemic circulation is supplied by this mixed blood.

One third of the TAPVD cases are in the cardiac type group in which left and right pulmonary veins make a common trunk behind the heart. Usually, this common trunk opens into the coronary sinus at the atrioventricular sulcus or sometimes directly into the right atrium[1,2].

With the infracardiac type of TAPVD, the pulmonary veins drain into the portal vein or its tributaries the ductus venosus, the inferior vena cava or the hepatic veins. The incidence of subdiaphragmatic abnormalities such as asplenia and polysplenia is not insignificant in this type of TAPVD[1,2].

During the infant period, 60% of the TAPVD cases have cyanosis starting in the first month, the rest of the patients present at medical centers with the symptoms and signs of congestive heart failure. In the first year 75% - 80% of the TAPVD cases die of severe pulmonary hypertension and congestive heart failure. The incidence of pulmonary hypertension is higher among the infracardiac TAPVD cases, and only a small proportion of these patients, with slowly developing pulmonary vascular disease, are likely to reach a more advanced age like our cases. Thus today, total correction of TAPVD is preferred during the neonatal or infant period[9,10].

When examining for TAPVD, mild or moderate cyanosis, a systolic murmur along the left sternal edge, fixed splitting of the second heart sound and the "snowman" sign on the chest X-ray contribute to the diagnosis (Figure 1). Usually, cardiac catheterization and echocardiography are essential to confirm the diagnosis of TAPVD.

All of our patients were functionally in NYHA class II, and only two of them had mild cyanosis. The mixing of pulmonary and systemic venous blood in TAPVD leads to O_2 saturation of 80% of the central venous blood. Thus, in TAPVD cases, cynosis is usually mild unless the ASD is large and the patient has severe pulmonary hypertension[2].

To date various surgical techniques have been performed in correction of TAPVD. In the past, hypothermia and inflow occlusion techniques were preferred. Also, in the first partial correction of TAPVD, a left posterolateral thoracotomy was carried out, and also extracardiac anastomosis between the common pulmonary sac and the left atrium using a closed technique[11]. Recently the majority of surgeons use a median sternotomy as the method of exposure for all types of TAPVD.

Most surgeons now agree that profound hypothermia with total circulatory arrest is particularly important in the repair of TAPVD in infants. During childhood too, the most popular method recently has been a cardiopulmonary bypass with moderate hypothermia of 26 - 28 °C and anoxic cardiac arrest, in fact, the type of operation we have been using. Further, the combining of this technique with cold K^+ cardioplegia and topical myocardial cooling (surface cooling) is believed to lead to better surgical results in the repair of TAPVD[11].

In the repair of supracardiac TAPVD we favor the transatrial approach, which was first described by Cooley and Ochsner[11] in 1957 and modified by Shumacher[12,13]. The advantages of this technique are a good exposure and the chance of performing a large anastomosis between the common pulmonary sac and the left atrium under direct vision. The elevation of the cardiac apex and the extracardiac approach for the anastomosis create certain difficulties related to the limited place in which to work. Some surgeons also use a left atrial appendix during these operations. Moreover, by this modality, the diameter of the anastomotic ring may be narrowed and this can lead to postoperative pulmonary congestion, as was seen in one of our cases, the one with the fatal outcome[11].

In the cardiac type of TAPVD cases, the pulmonary veins usually open into the large coronary sinus. In this condition, the orifice of the coronary sinus is enlarged close to the left atrial wall to establish a communication with ASD which is generally coexistent. Then closure of ASD is carried out with a large pericardial or teflon patch, so that the drainage of the pulmonary veins via the enlarged orifice of the coronary sinus is directed to the left atrium[14]. As we have seen in one of our cases, it is not usual to open all the pulmonary veins directly into the right atrium, in the cardiac type of TAPVD[1,2]. In our case, we closed the ASD with a pericardial patch in order to direct the drainage of the pulmonary veins into the left atrium. We also made an incision towards the left upper pulmonary vein on the atrial septal remnant. We believe the addition of this modality will also improve the drainage of the pulmonary veins into the left atrium.

The diameter of the anastomosis between the common pulmonary sac and the left atrium is one of the most important determinants affecting postoperative mortality and morbidity particularly in the supracardiac type of TAPVD[11]. In neonates and infants it is unlikely that a satisfactory passage will be reached each time. Therefore with this age group it has been suggested the anastomosis be

made as large as possible and that interrupted sutures be used so that it may be enlarged at a later date[11,15].

The development at an early stage of pulmonary vascular disease affects postoperative mortality as much as does surgical technique[11,16].

The necessity of left atrial enlargement and the effect of a small left atrium are still controversial subjects[11,17]. It is quite possible to enlarge the left atrium by deviation of the septum towards the right atrium with a patch when the left atrial cavity is assessed as small for the patient. However, it has been reported that there is no significant difference in survival rates for those with and those without left atrial enlargement in the repair of TAPVD. This may be because efforts to improve the size of the left atrium by surgically enlarging it are not very effective and because the lack of left atrial reservoir function is rarely critical if all other aspects of the situation are favorable to survival[11].

As indicated earlier, the development of pulmonary congestion due to anastomotic stricture or a small anastomotic ring, is an important factor behind delayed mortality[11,18]. This is particularly important in the repair of TAPVD during the infant period. Thus when there is not satisfactory interatrial mixing, it is advisable to first perform a balloon atrial septostomy, to give the baby the chance of a quiet infant period and delay corrective surgery till the patient is two to three years of age[18].

In conclusion, while the overall mortality rate is about 80% for the natural prognosis, excellent results can be achieved by total correction of TAPVD.

Summary

In this paper, we report five cases of TAPVD all of which underwent total correction. We suggest that total correction by the transatrial approach is the treatment of choice in cases of TAPVD.

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