SURGICAL TREATMENT OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

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ABSTRACT

Fourteen infants ranging in age from 18 days to 10 months (median age = 4 months) underwent surgical repair of total anomalous pulmonary venous connection (TAPVC) between September, 1988 and December, 1989. The anomalous drainage was supracardiac in nine, cardiac in two, infracardiac in one and mixed in one. One patient had a complex type of TAPVC. There were two hospital deaths, one a critically ill three week old infant with obstructed infracardiac TAPVC and the other with complex TAPVC. There was no late death. All twelve survivors are in NYHA functional Class 1. Postoperative echocardiography revealed unobstructed pulmonary blood flow in all of them. Surgical correction of TAPVC in infancy has been performed with gratifying results. A high index of suspicion, early diagnosis, prompt referral and an aggressive surgical approach are essential for success in this otherwise lethal condition.

Key words: *Anomalous pulmonary venous connection, Infancy, Phenoxybenzamine.*

Corrective operation for total anomalous pulmonary venous connection (TAPVC) has become increasingly successful during the past decade, mainly because of the widening experience with intraoperative management of very small infants(1-3). However, the surgical mortality still remains as high as 8 to 20% in most series(4-7).

Surgical correction of TAPVC in infancy has not been reported so far in the Indian literature. This report describes our initial experience with surgery for TAPVC in infancy.

Material and Methods

Fourteen patients, 18 days to 10 months age (median age = 4 months) underwent operation for TAPVC from September, 1988 to December, 1989. The anomalous drainage was supracardiac (to the superior vena cava via the innominate vein) in 9 patients (*Fig. 1*), cardiac in 2 patients (one via the coronary sinus, 1 via multiple openings in the right atrium), infracardiac in 1 patient and mixed (supracardiac and cardiac) in 1 patient. One patient had complex TAPVC associated with ventricular septal defect and hypoplastic right pulmonary artery (*Figs. 2a and 2b*).

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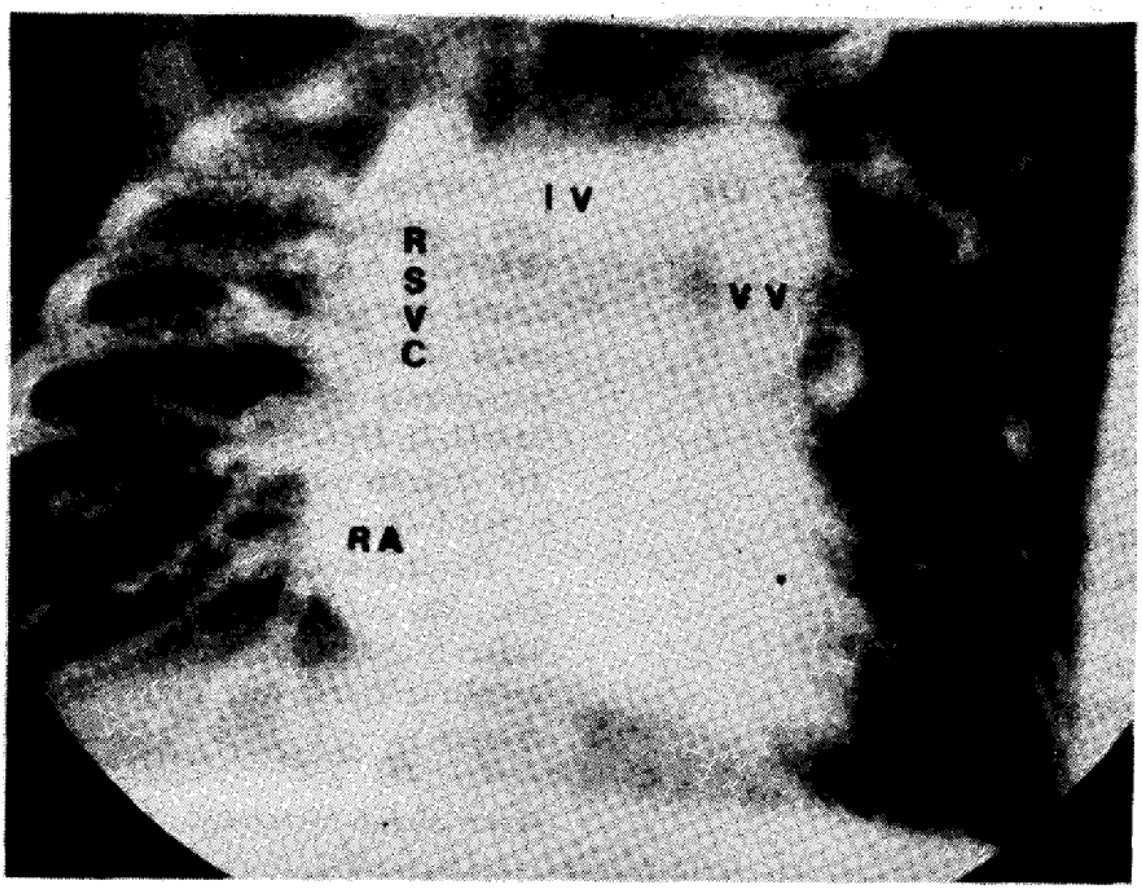

Fig. 1. Angiogram showing classical supracardiac type of TAPVC.

The majority of patients were symptomatic. There was cyanosis in 4, poor feeding and failure to thrive in 6 and persistent congestive heart failure in 5. Two patients were in extremis and required preoperative ventilation and inotropic support.

Preoperatively all patients were evaluated by two dimensional and doppler echocardiography and 7 patients additionally by cardiac catheterization and angiography. In these seven patients the pulmonary artery peak systolic pressure ranged between 26 and 120 mm Hg (median = 55 mm Hg). The mean pulmonary artery pressure was between 16 and 80 mm Hg (median = 35 mm Hg). Evidence of pulmonary venous obstruction was present in three patients.

Surgical Treatment

All patients were operated under cardiopulmonary bypass and profound hypothermia (18-20° C). Cold blood cardioplegia was used for myocardial protection. Phenoxybenzamine was administered to all patients for alpha adrenergic blockade just before cardiopulmonary bypass was begun. The ductus (or ligamentum) arteriosum was routinely dissected and ligated during the cooling phase. Intermittent periods of low flow bypass and total circulatory arrest were used to facilitate the operative procedure as and when required.

In all cases of supracardiac and infracardiac TAPVC, the apex of the heart was elevated over to the right side and an anastomosis was constructed between the left atrium and common pulmonary vein(8).

The anastomosis was made as large as possible. An approach via the right atrium was used for repair of the cardiac type of TAPVC, suturing a Gortex (Expanded

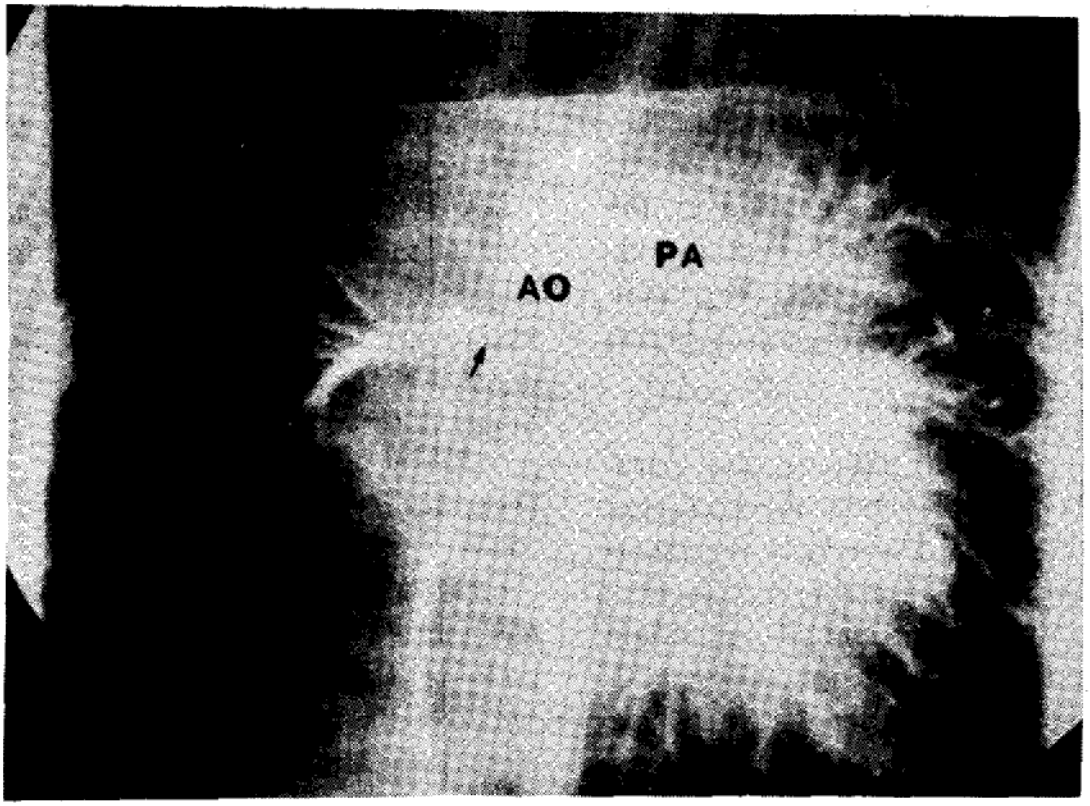

Fig. 2a. Angiogram with right ventricular injection filling both aorta (AO) and pulmonary arteries, due to presence of VSD. Note hypoplastic right pulmonary artery (arrow).

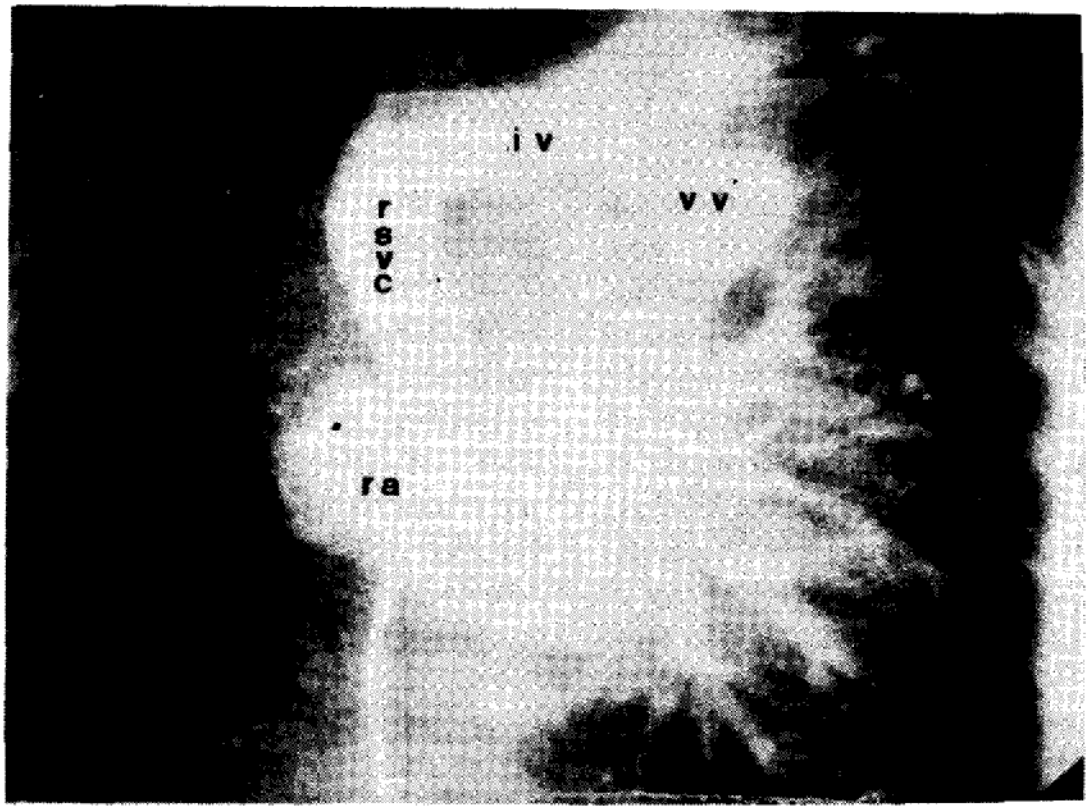

Fig. 2b. Levo phase of right ventricular angiogram of same patient showing supracardiac type of anomalous drainage of left sided pulmonary veins. (VV = vertical vein, IV = innominate vein, RSVC = Right superior vena cava, RA = atrium).

Polytetrafluroethylene) patch so as to direct the pulmonary venous blood through a surgically created large atrial septal defect into the left atrium. TAPVC of the coronary sinus type was repaired by the method of Van Praagh *et al.*(9) without using any prosthetic material. Enlargement of the left atrium was not necessary in any patient.

Postoperatively all patients were paralysed and the lungs mechanically ventilated for atleast 24 hours. Pulmonary hypertension was managed by a regimen of moderate hyperventilation (aiming to maintain arterial carbon dioxide tension between 30-35 mm Hg) and alpha adrenergic blockade with phenoxybenzamine in all patients according to individual patient responses. Additional vasodilators like sodium nitroprusside and nitroglycerine were sometimes necessary for a breakthrough in pulmonary hypertensive crisis.

Results

There were two hospital deaths. One patient with a complex TAPVC who underwent correction along with VSD closure showed signs of critically depressed cardiac function and could not be weaned off cardiopulmonary bypass. The second patient was a three week old critically ill infant with obstructed infracardiac TAPVC with a pulmonary artery peak systolic pressure of 120 mm Hg and a mean pressure of 80 mm Hg. The patient continued to have suprasystemic pulmonary artery pressure after being weaned from cardiopulmonary bypass, despite an adequate surgical repair. The child died of right ventricular failure during a pulmonary hypertensive crisis. The 12 survivors have been followed up clinically from 4 months to 19 months (mean=9 months) postoperatively. All remain clinically well. Antifailure medications were tapered of in all patients within six weeks of surgery.

A postoperative two dimensional Doppler echocardiography was performed in all 12 survivors and showed nonobstructive pulmonary venous flow into the left atrium in all of them. There was no residual shunt in any of them.

Discussion

TAPVC usually occurs as an isolated congenital cardiac malformation. The majority of patients with TAPVC have severe symptoms, with some in extremis and only 20% patients survive the first year of life(10). Surgical repair offers the only chance of survival. Among the 13 patients with isolated TAPVC in our series 12 survived, a hospital mortality rate of 7.7% which compares favorably with the figures reported in the recent literature(4-7). However, the hospital mortality rate can still be high in neonates with pulmonary venous obstruction who are in poor preoperative condition and in those with complex TAPVC(11,12).

Patients with TAPVC have a tendency to develop obstruction to pulmonary venous drainage and then deteriorate rapidly. This also causes an earlier onset of pulmonary hypertension and can thus increase the risk of surgery. Thus a high index of suspicion and early referral are important to reduce mortality. Further we believe like others(13) that noninvasive preoperative diagnosis in contrast to invasive diagnosis allows the patient to be in a better condition for operation.

Intimal hypertrophy and fibrosis of the pulmonary vessels are a common finding in patients with TAPVC and these vessels

are capable of further vasoconstriction in response to stimuli such as infection, hypoxia, hypercarbia and acidosis. For this reason careful attention to postoperative acid-base balance and ventilatory support during paralysis is required, in order to prevent pulmonary hypertensive crisis.

The posterior approach to the left atrium as described by Williams *et al.*(8) provides excellent exposure and allows creation of a wide anastomosis without any problems with orientation or twisting of the suture line. Our results support the contention that it is rare for the left ventricle or atrium to be too small to support the systemic circulation. Two dimensional echocardiography showed unobstructed pulmonary venous flow in all survivors. It is likely that a continuous suture technique is adequate for an unobstructed anansto-mosis even in neonates.

It is concluded that isolated TAPVC usually presents during the first months of life and demands urgent surgical intervention, especially when the pulmonary venous flow is obstructed. With recent advances in cardiac surgery the incremental risk of younger age at operation is being progressively neutralized. Presence of pulmonary venous obstruction can increase the risk of operation. However, its presence dose not indicate conservative medical management in view of the poor natural history of the disease and the very good survival rate after aggressive surgical treatment. Further, the late results of the operation are usually gratifying and the need for reoperation seems unlikely. Thus a high index of suspicion, use of echocardiography for an early accurate diagnosis and early referral for surgery are all important to reduce the mortality of this otherwise lethal condition.

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NOTES AND NEWS

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